Virchows Archiv (1994) 424:217–224

CASE REPORT

Leif Johansson · Birgitta Carlén

Sarcoma of the pulmonary artery: report of four cases with electron microscopic and immunohistochemical examinations, and review of the literature

Received: 20 October 1993 / Accepted: 17 November 1993

Abstract Herein we report the clinicopathological features of four cases of pulmonary artery sarcoma that appeared at our institution during a period of 30 years. The patients, 2 males and 2 females, were 50–62 years old. Tumour was found in the pulmonary trunk and right pulmonary artery in all cases, in the pulmonary valve and left pulmonary artery in three of the four cases, and in the right ventricular outflow tract in one case. There was direct extension or metastases to the lungs in two cases, the heart in one case, mediastinum or lymph nodes in two cases and the pleura in one case. Ultrastructural examination in one case revealed cells with features of smooth muscle cells and myofibroblasts. Immunohistochemical examination of three cases gave the following results: vimentin and smooth muscle specific actin was positive in all three cases, desmin in one case and cytokeratin in one case. No positivity was found for Factor VIII. This and other studies indicate that histologically most pulmonary artery sarcomas are leiomyosarcomas or "undifferentiated spindle cell sarcomas". Immunohistochemical and ultrastructural examinations favour an origin from myofibroblasts, probably derived from multipotent (undifferentiated) cells in the wall of the vessel. Most lesions show extensive intrathoracic growth although they rarely metastasize outside the thoracic cavity. They have a poor prognosis although some cases are currently being diagnosed during life.

Key words Pulmonary artery · Neoplasm · Sarcoma Immunohistochemistry · Electron microscopy

L. Johansson (✉) · B. Carlén
Department of Pathology, University Hospital, S–221 85 Lund, Sweden

Introduction

Sarcomas of the pulmonary artery (PAS) are usually primary in the pulmonary trunk, but may also be located in the main pulmonary arteries, the pulmonary valve and the right ventricular outflow tract. The first case was reported by Mandelstam in 1923. Since then case reports as well as several reviews have appeared (Bleisch and Kraus 1980; Baker and Goodwin 1985; Nonomura et al. 1988; McGlennen et al. 1989; Burke and Virmani 1993). In 1980 Bleisch and Kraus reported 60 cases, 28 of which were from the non-English literature. Baker and Goodwin reviewed the English literature in 1985 and found 45 cases, and in 1988 Nonomura et al. were able to collect 110 cases from the world literature. Apart from one radiation-induced case (Shah et al. 1991) there are no known causes of PAS. Preoperative diagnosis by means of computed tomography (CT) and magnetic resonance imaging have been reported (Smith et al. 1989), and there are reports of successful treatment with a combination of resection and homograft reconstruction (Dossche et al. 1992; Head et al. 1992; Okada et al. 1993).

Most histopathological types of sarcoma have been reported, including tumours with differentiation towards chondroid or bone tissue (Nonomura et al. 1988). With some exceptions (Paulsen and Egeblad 1983; Nonomura et al. 1988; McGlennen et al. 1989), electron microscopic examinations have been carried out with poorly preserved autopsy material. Immunohistochemical findings have been reported in 18 cases altogether (Nonomura et al. 1988; McGlennen et al. 1989; Kaiser et al. 1990; Nerlich et al. 1990; Hiroshima et al. 1992; Ramp et al. 1992; Burke and Virmani 1993).

In this report we add four cases from the files of the Department of Pathology, University Hospital, Lund, Sweden. In three cases material for immunohistochemical examinations was available. In one of these cases material from a surgical biopsy was also available for electron microscopic examination. The literature is summarized concerning the gross morphological, histo-

pathological and immunohistochemical features of these exceedingly uncommon tumours.

Case reports

An autopsy was performed in all cases: in cases 1 and 2 at the local hospitals to which the patients were admitted, and in cases 3 and 4 at the Department of Pathology, University hospital, Lund.

Case 1 (1961)

A 60-year-old female was admitted to hospital with cyanosis but without severe dyspnoea. Pulmonary thromboembolism was suspected and she received anticoagulant therapy. However, she deteriorated fast and died rapidly. A tumour the size of the end of a small finger was found on one of the semilunar cusps of the pulmonary valve and the main part of the pulmonary trunk was occupied by a tumour, 3 cm in diameter, that was hard and lobulated, had a chondroid cut surface and extended into both of the pulmonary arteries. Histologically the tumour was composed of slightly atypical chondroid tissue with formation of osteoid and fibrous tissue with a varying degree of atypia. Few mitoses were found. A low-grade chondrosarcoma was suggested.

Case 2 (1964)

A formerly healthy 62-year-old male was admitted to hospital in March 1963 with pneumonia and an abscess in the left upper lobe. Chest radiography suggested cor pulmonale. He developed dyspnoea and oedema of the feet and suffered from nausea and vomiting. He subsequently died in August 1964, about 1.5 years after the first admission to hospital, without even a tentative diagnosis. The heart and lungs were sent en bloc to our laboratory for histopathological examination. Gross morphological examination showed dilatation and hypertrophy of the right ventricle. The pulmonary trunk was filled with fragile, partly necrotic tumour masses that infiltrated through the wall of the vessel and extended into both of the pulmonary arteries. There were small, peripheral infarcts in the lungs and metastases in the mediastinal lymph nodes. Histological examination revealed a spindle cell sarcoma that grew in storiform and whorled patterns. Multinucleated giant cells were found in some areas. Also, hyalinization and necrosis were found. The tumour was designated as a fibrosarcoma.

Case 3 (1986)

A 48-year-old male presented with dyspnoea in 1984. From that time on he received anticoagulant therapy for "chronic pulmonary embolism". However, other diagnoses, such as fibrosing mediastinitis and malignancy, were also considered. In the summer of 1986 an angiography showed total occlusion of the right branch of the pulmonary artery and occlusion of some minor intrapulmonary branches on the left side. As the findings were in accordance with the initial diagnosis the anticoagulant therapy was sustained, together with corticosteroids. Due to suspected vasculitis he was also treated with immunosuppressive drugs. In November 1986 severe arrhythmias developed. By then he had severe dyspnoea; he deteriorated fast and died about 1 month after final admission to hospital. At autopsy, tumour nodules were found on two of the semilunar cusps of the pulmonary valve and in the pulmonary trunk. There was also total occlusion of the right, and a partial occlusion of the left, pulmonary artery. The tumour grew through and destroyed the walls of the vessels and invaded the lungs and pleura. Lung infarctions were present bilaterally but were more pronounced on the right side. Histological examination showed a tumour composed of spindle cells with scattered areas of pronounced atypia. It was sometimes difficult to separate tumour from connective tissue, especially in the intrapulmonary branches of the pulmonary artery. Large areas were necrotic, however, and sometimes replaced by reactive fibrous tissue. A spindle cell sarcoma, possibly leiomyosarcoma, was suggested.

Case 4 (1988)

A 60-year-old female presented with a history of increasing dyspnoea for 6 months and right-sided chest pain. On admission to hospital she had signs of pleuritis, cough, blood tinged sputum and fever. Chest radiography showed infiltration of the right middle lobe and echo-cardiography revealed a smaller rounded tumour at the outflow tract of the right ventricle. A pressure gradient of 20 mmHg, impaired function of the right ventricle and insufficiency of the tricuspid valve was also found. A thoracotomy revealed a tumour that involved one of the cusps of the pulmonary valve, the pulmonary trunk and right pulmonary artery. Resected tumour consisted of several large white to yellow, firm fragments up to 3 cm diameter with a smooth and glistening surface. Frozen section was consistent with a sarcoma. The patient had a cardiac arrest and died during surgery. At autopsy the base of the tumour was found on one of the cusps of the pulmonary valve and the immediate infracuspid region, attached to a 3×3 cm large area. The tumour extended through the wall of the pulmonary trunk, infiltrated the surrounding tissue, grew over the aorta and displaced the right coronary artery. No tumour was found in the left lung or the left pulmonary artery. A lymph node metastasis was found in the mediastinum. Microscopical examination revealed a tumour composed of spindle cells with elongated nuclei, sometimes in a fascicular pattern, with a varying degree of atypia. Cells reminiscent of fibroblasts as well as multinucleated giant tumour cells were encountered. The tumour grew through the wall of the pulmonary trunk and infiltrated the surrounding tissue, including the heart. Necrosis and areas with myxoid ground substance were seen. The tumour was considered to be a leiomyosarcoma.

Table 1 summarizes the results of the gross morphological and microscopical examination. Thus, the tumours involved the pulmonary trunk and the right pulmonary artery in all cases, and the left pulmonary artery in three. In addition, tumour infiltrated the cusps of the pulmonary valve in three of the cases but the right ventricular outflow tract was involved in only one case. One tumour was initially designated as a low-grade chondrosarcoma while the other three were considered variants of spindle cell sarcomas.

Materials and methods

For electron microscopy (case 4) fresh tissue was fixed in 2% glutaraldehyde in 0.1 M sodium cacodylate buffer, pH 7.2, post-fixed in 2% osmium tetroxide in *s*-collidine buffer, dehydrated in ethanol, transferred to propylenoxide and embedded in agar resin 100. Ultrathin sections were cut on a LKB Ultrotome III, counterstained with uranyl acetate and lead citrate. The sections were examined at 60 kV in a Zeiss EM 10 electron microscope.

For immunohistochemistry (cases 1, 3, 4) a panel of monoclonal antibodies (Table 2) was used with formalin-fixed and paraffin-embedded tissue. The sections were deparaffinized and incubated with 0.5% hydrogen peroxidase (H_2O_2) in methanol for 30 min to block the endogenous peroxidase activity. All sections were pretreated with 0.1% trypsin in 0.1% calcium chloride solution (pH 7.8) at 37° C for 15 min. Immunohistochemical labelling was performed with the indirect method with the primary antibody incubated 1 h at room temperature. The staining reaction was developed with 0.025% 3.3 diaminobenzidine-tetrahydrochloride and 0.01% H_2O_2 for 5 min and counterstained with Mayer's haematoxylin.

Table 1 Gross morphological findings in four cases of sarcoma of the pulmonary artery (*RVOT* right ventricular outflow tract, *VP* pulmonary valve, *TP* pulmonary trunk, *APd* right pulmonary artery, *APs* left pulmonary artery, *L* lung, *H* heart, *M* mediastinum, *LN* lymph node, + present, o absent)

Case no.	Sex/age	Location of tumour					Direct extension			Metastasis			Diagnosis
		RVOT	VP	TP	APd	APs	L	H	M	L	LN	Pleura	
1	F/60	0	+	+	+	+	0	0	0	0	0	0	Chondrosarcoma
2	M/62	0	0	+	+	+	0	0	+	0	+	0	Fibrosarcoma
3	M/50	0	+	+	+	+	+	0	0	+	0	+	Leiomyosarcoma
4	F/60	+	+	+	+	0	+	+	0	0	+	0	Leiomyosarcoma

Table 2 Antibody panel

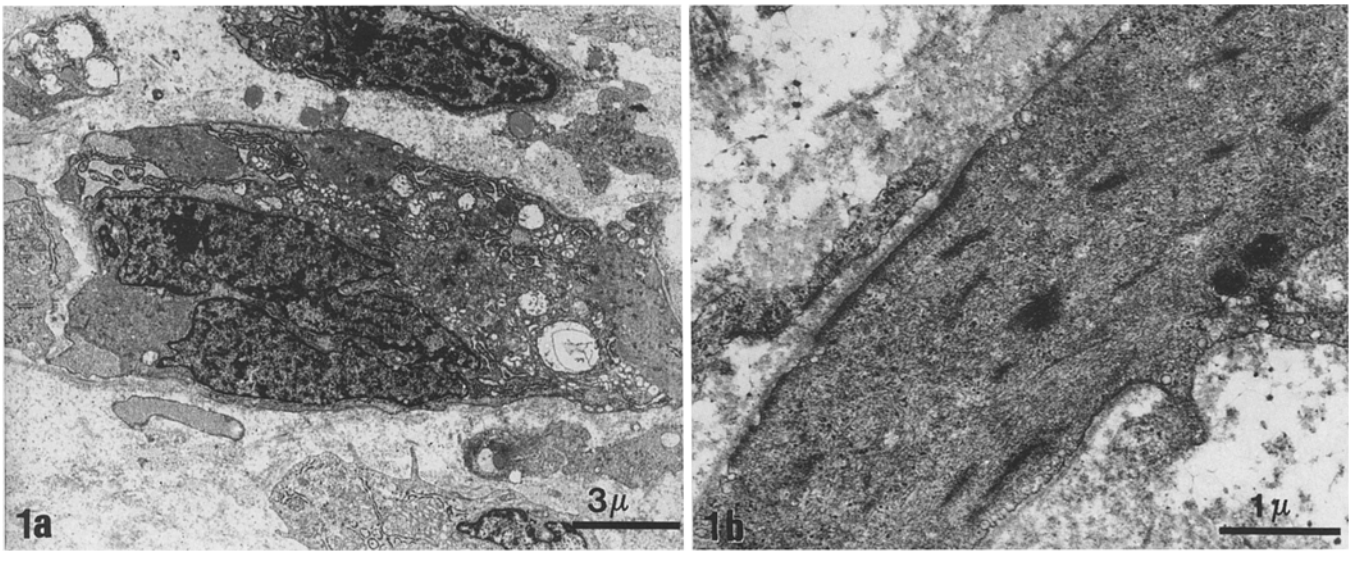
Antibody	Source	Dilution
Cytokeratin AE1/AE3	Hybritechs, San Diego, Calif., USA	1:400
Cytokeratin CAM 5,2	Becton and Dickinson, Calif., USA	1:5
Vimentin	DAKO, Copenhagen, Denmark	1:10
Desmin	DAKO	1:300
Smooth muscle specific actin	DAKO	1:500
Factor VIII	DAKO	1:1000 (overnight)

Results

On electron microscopy (case 4) the most frequently occurring cell type had features indicating a smooth muscle origin (Erlandsson 1981; Mackay 1987: Ghadially 1988; Fisher 1990) with most nuclei being large and oval with irregular or indented nuclear membranes and nuclear chromatin randomly dispersed. The cells contained thin actin filaments with focal densities distributed parallel to the nuclei or along the plasma membranes, numerous micropinocytotic vesicles and subplasmalemmal densities alongside and near the plasma membrane. Other intracytoplasmic structures were mitochondria, free ribosomes, abundant rough endoplasmatic reticulum, a few lipid droplets and glycogen. An external lamina tapered along the outside of the plasma membrane (Fig. 1). A less frequently occurring cell type was elongated, contained peripherally located bundles of myofilaments with focal densities, prominent dilated rough endoplasmatic reticulum and Golgi apparatus. Occasional micropinocytotic vesicles were seen along the plasma membrane. The appearance was consistent with myofibroblasts (Fig. 2; Eyden 1991).

Immunohistochemical findings are outlined in Table 3. In case 1 scattered spindle cells reacted with both of the cytokeratins. The reaction was distinct, although relatively weak, with cytokeratin AE1/AE3 (Fig. 3) while only scattered cells reacted with cytokeratin CAM 5.2. Vimentin gave a strong reaction with all the tumours (Fig. 4). Smooth muscle specific actin reacted strongly with the spindle cell component in all tumours

Fig. 1a, b Case 4. **a** Leiomyocyte with markedly cleaved nucleus. Original magnification ×3,900. **b** Portion of neoplastic leiomyocyte showing actin microfilaments with fusiform dense bodies, attachment plaques, pinocytotic vesicles and continuous external lamina. Original magnification ×15,500



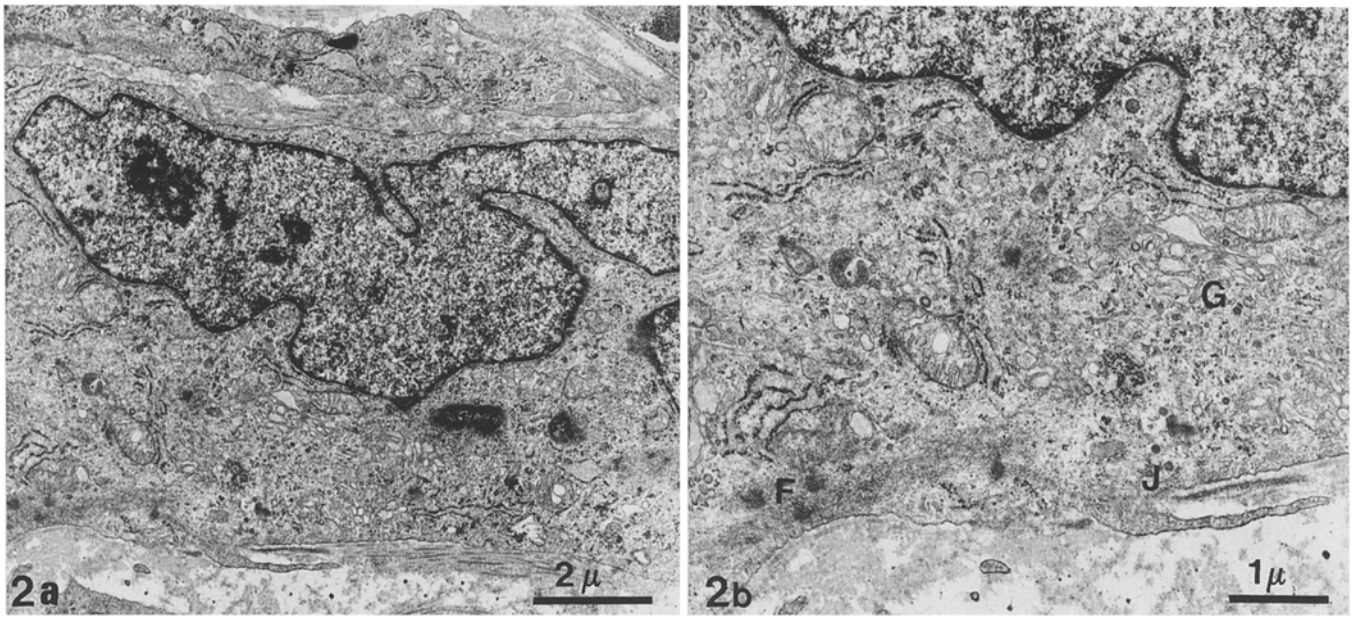


Fig. 2a, b Case 4. **a** Myofibroblast. Original magnification × 6,610. **b** Detail showing subplasmalemmal actin microfilaments with focal densities (*F*), Golgi apparatus (*G*), endoplasmic reticulum and at the cell surface a fibronexus junction (*J*). Original magnification × 11,500

Table 3 Immunohistochemical findings in 21 pulmonary artery sarcomas. Three of our cases and 18 cases reported in the literature (*MS* muscle specific, *SMS* smooth muscle specific, *ND* not done)

Author	Cytokeratin	Vimentin	Desmin	Actin		Factor VIII	Diagnosis
				MS	SMS		
Nonomura (1988)							
Case 1	ND	ND	–	+	ND	–	Malignant mesenchymoma
Case 2	–	ND	–	+	ND	–	Malignant fibrous histiocytoma
McGlennen (1989)							
Case 1	–	+	–	+	ND	–	Osteosarcoma
Case 2	–	+	–	+	ND	–	Chondrosarcoma
Case 3	–	+	–	–	ND	–	Spindle cell sarcoma
Case 4	–	+	–	+	ND	–	Spindle cell sarcoma
Kaiser (1990)	–	+	–	–	ND	–	Malignant fibrous histiocytoma
Nerlich (1990)	–	ND	+	ND	ND	ND	Leiomyosarcoma
Hiroshima (1992)	–	+	–	–	–	ND	Undifferentiated sarcoma
Ramp (1992)							
Case 1	–	+	–		–	ND	Fibrosarcoma
Case 2	–	+	–		–	ND	Malignant fibrous histiocytoma
Burke (1993)							
5 cases	–	+	–	ND	+	–	"Intimal sarcoma" (5 cases)
2 cases	–	+	+	ND	+	–	"Leiomyosarcoma" (2 cases)
Johansson (1993)							
Case 1	+	+	–		+	–	
Case 3	–	+	–		+	–	
Case 4	–	+	+		+	–	

(Fig. 5). Desmin was strongly and distinctly positive only in case 4 (Fig. 6). Factor VIII was negative in all tumour cells but invariably positive with the endothelium of the vessels, in the tumours, as well as in normal tissue.

Discussion

It is reasonable that early detection of PAS may influence the prognosis and extensive intraluminal growth may often give rise to early symptoms. Despite this, the

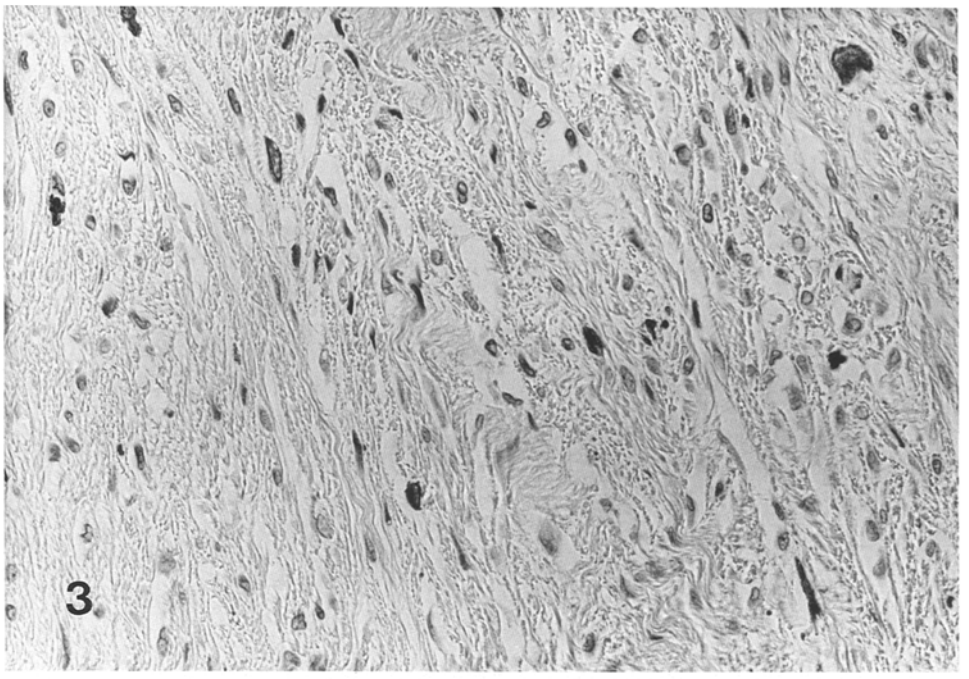


Fig. 3 Case 1. Several spindle cells reacts with cytokeratin AE1/AE3, either in long slender cytoplasmic processes (*right lower corner*) or perinuclear. ×400

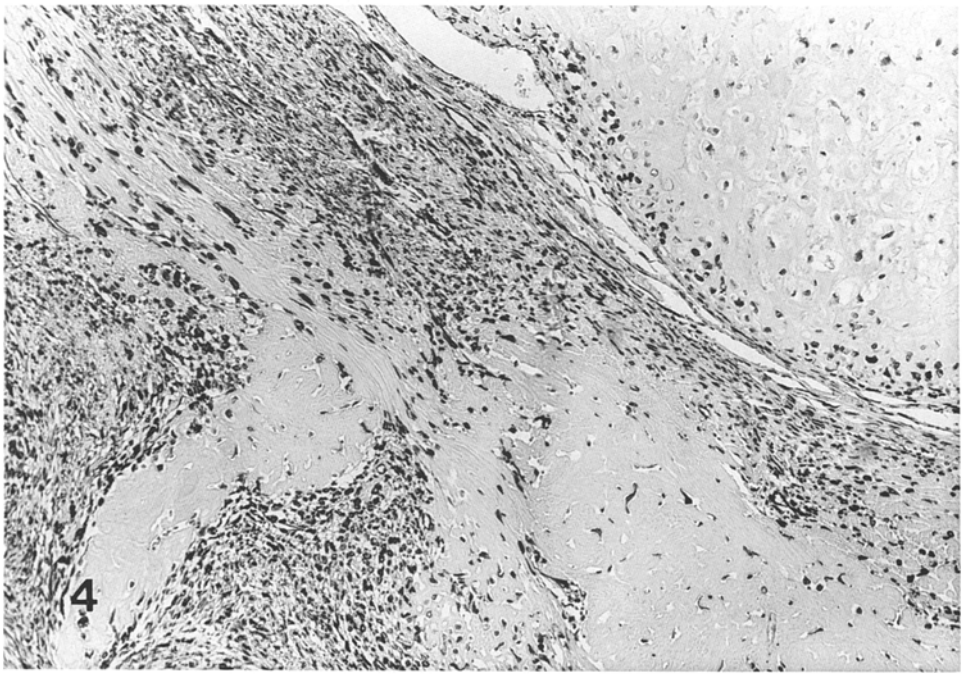


Fig. 4 Case 1. Tumour cells show a strong reaction with vimentin. Note relatively mature chondroid tissue at upper right corner. ×100

diagnosis is often delayed, as in our cases 1 and 3, with clinical presentation as "chronic pulmonary embolism" (Burke and Virmani 1993). With the possible exception of those tumours containing bony and chondroid tissue (see below) no histological subgroup of PAS has a more favourable prognosis. Even if evolution of the symptoms is quite insidious, the tumours are usually overtly malignant and the expected survival from diagnosis is 12–24 months (McGlennen et al. 1989). However, preoperative diagnosis by means of CT and magnetic resonance imaging has been reported (Smith et al. 1989) and as distant metastasis is a rare, or a late occurring, event, PAS may be amenable to surgical invention. Successful treatment with a combination of resection and homograft reconstruction (McGlennen et al. 1989; Dossche et al. 1992; Head et al. 1992; Okada et al. 1993), as well as prolonged survival with the use of adjuvant chemotherapy (Head et al. 1992), has been reported.

Bony and cartilaginous tumours of the heart and great vessels, such as the first of our cases, may be regarded as a subgroup with special clinicopathological features. The patients are usually women in their fourth or fifth decade. They arise at the base of the heart (mostly in the pulmonary artery but also in the atria or mitral

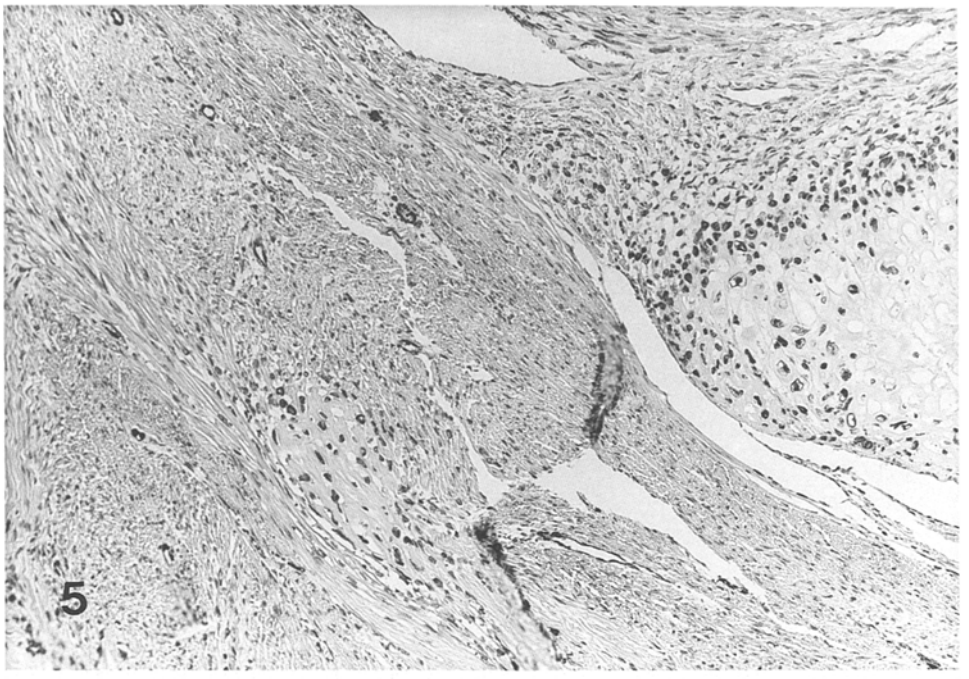


Fig. 5 Case 1. Tumour cells react strongly with smooth muscle specific actin. Chondroid tissue to the right. ×100

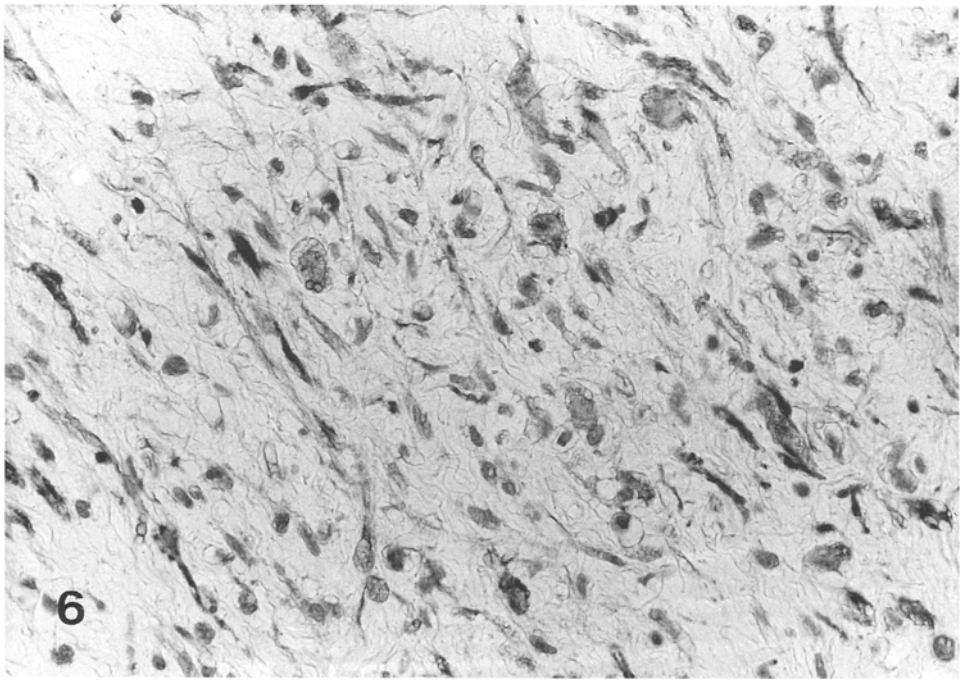


Fig. 6 Case 4. Virtually all cells shows a positive reaction with desmin. ×400

valve) and are usually slowly aggressive (McConnell 1970; Johansson et al. 1989) but eventually fatal because of their location. All contain a spindle cell component in addition to chondroid or bony tissue and histologically they are often designated as chondrosarcomas, osteosarcomas or malignant mesenchymomas. The formation of bone and chondroid tissue is thought to occur through metaplasia in a fibrosarcoma or another fibrous tumour (McConnell 1970). Bony and cartilaginous tumours are said not to occur in other large vessels (aorta and vena cava; Burke and Virmani 1993).

By definition, the site of origin of PAS is the various parts of arteria pulmonalis, the pulmonary valves, and the right ventricular outflow tract. Often the tumour is loosely attached to the wall of the vessel and grows with polypoid intraluminal extensions and with only focal invasion of the wall of the vessel (McGlennen et al. 1989). Nonomura et al. (1988) collected 110 cases, including two of their own from the world literature, and found that the pulmonary trunk was involved in 80% of cases, the right and left pulmonary arteries in about 60% each, both pulmonary arteries in about 40%, the

pulmonary valve in 30% and the right ventricular outflow tract in 8%. There were metastatic lesions to, or direct invasion of, the lungs in 40% and mediastinal tissue in 11% of the cases with distant metastases in 19%. The figures concerning site of origin roughly correspond to our findings (Table 1), except that the pulmonary valve was comparatively frequently involved in our cases (3 out of 4 compared to 30%). We found direct extension to the lungs in two cases, to the heart in one case and to the mediastinum in one case. Metastatic lesions were found in the lungs in one case, lymph nodes in one case and pleura in one case. Metastases were not found outside the thoracic cavity despite autopsy examination of all our cases.

In our four cases there were two "undifferentiated spindle cell sarcomas", one low-grade chondrosarcoma and one leiomyosarcoma. Nonomura et al. (1988), in their review of 110 cases of PAS, found 34 "undifferentiated sarcomas", 23 leiomyosarcoma, three "myogenous sarcoma", 7 each of rhabdomyosarcoma, fibrosarcoma and "malignant mesenchymoma". There were also 10 cases of myxo- or fibromyxosarcomas, 4 chondrosarcomas, 3 osteosarcomas, 4 pure angiosarcomas and 3 tumours with angiosarcomatous differentiation in combination with other components. Finally, there were 3 malignant fibrous histiocytomas and 1 liposarcoma.

We found a strong or moderately strong reaction for vimentin and smooth muscle specific actin in our three cases (Table 3). Immunohistochemical findings have previously been reported in 18 cases, all of which were positive with vimentin. In one study, staining for smooth muscle specific actin was positive in six out of seven tumours (Burke and Virmani 1993; Table 3) while in three other cases it was negative (Hiroshima et al. 1992; Ramp et al. 1992; Table 3). Dual expression of vimentin and smooth muscle specific actin is said to be characteristic for myofibroblasts and/or myofibroblastic tumours (Rangdaeng and Truong 1991; Burke and Virmani 1993). Myofibroblasts are cells with features of both smooth muscle cells and fibroblasts that constitute the cellular part of normal scar tissue and seem to occur in tumours or tumorous lesion that contain fibroblasts. It has been doubted whether a pure fibroblastic lesion really exists (Lundgren 1993).

The issue is more complicated, however. Cytokeratin staining has invariably been negative in previous immunohistochemical studies (Table 3). Surprisingly, in our case 1, regarded as a low-grade chondrosarcoma, scattered spindle cells were immunoreactive with the cytokeratins AE1/AE3 and CAM 5.2. In a previous article concerning cardiac myxomas dual expression of CAM 5.2 and vimentin in a group of myxoma cells without a glandular arrangement was reported (Johansson 1989). It was then suggested that some myxoma cells have the same intermediate filament profile as activated myofibroblasts in pleura and peritoneum, generally known as reactive non-neoplastic subserosal cells, that express cytokeratin and vimentin (Bolen et al. 1986, personal observation). Thus, it is likely that the cytokeratin-positive spindle cells in case 1 were cells with differentiation towards myofibroblasts. Expression of cytokeratins in sarcomas and sarcomatous lesions with myofibroblastic differentiation is sometimes referred to as aberrant (Lundgren 1993). In reactive non-neoplastic subserosal cells immunoreactivity for cytokeratin is obligate, however, and these cells are certainly myofibroblasts ultrastructurally (Bolen et al. 1986). Also, an electron microscopic examination of case 4, regarded as a leiomyosarcoma, showed smooth muscle cells and myofibroblasts (Fig. 2). Ultrastructural features of myofibroblasts, usually in combination with other components, have previously been reported in PAS (Paulsen and Egeblad 1983; Nonomura et al. 1988; McGlennen et al. 1989). Thus, virtually all PAS, like many other soft tissue tumours, contain cells with myofibroblastic features and it may also be anticipated that some of these cells have the ability to express cytokeratin.

Tumour in case 4 of our study was positive with desmin (Table 3) and had cells with ultrastructural features of smooth muscle cells (Fig. 1). Desmin antibodies have been used in all previous immunohistochemical studies (Table 3); 3 of 18 showed a positive reaction and were thus regarded as leiomyosarcomas. This indicates that, if the diagnosis of leiomyosarcoma is concurrent with a positive reaction for desmin, only a minority of PAS can be regarded as leiomyosarcoma. On the one hand, the view that positive staining for desmin is equivalent with leiomyosarcoma in the absence of light microscopic features consistent with the diagnosis (that immunohistochemistry provides histogenetical information) has been challenged (Allen 1990) and it has also been shown that as many as 17% of nonmyogenic soft tissue tumours may express desmin (Rangdaeng and Truong 1991). On the other hand, many desmin antibodies lack specificity and it has been shown that considerably more cases will be positive if a panel of desmin antibodies are used (Lundgren 1993). In summary, to prove that a tumour is a leiomyosarcoma, in the pulmonary artery or other locations, more than one desmin antibody may have to be used, but histiogenesis should not be regarded as established unless light microscopy, and/or electron microscopy, is consistent with the diagnosis.

Muscle specific actin was positive in five out of eight reported cases (Table 3). This antibody, however, has the disadvantage of a very broad specificity and reacts with most kinds of muscle tissue. Hence, it is not suitable for differentiating leiomyosarcoma from other myogenic sarcomas. Factor VIII has been invariably negative in this and other immunohistochemical studies (Table 3), which corroborates the fact that Nonomura et al. (1988) found only 4 angiosarcomas among 110 reviewed cases of PAS.

As the entity PAS is mainly defined by clinical presentation and anatomical location, and as the histological forms are diverse, there is poorly defined ground for histogenetic considerations. However, with the aid of immunohistochemistry and electron microscopy, a myofibroblastic origin of PAS has been proposed (McGlennen et al. 1989) and it has also been speculated that

many PAS are derived from intimal smooth muscle cells or fibroblasts, either of which can be the ancestors of myofibroblasts (Burke and Virmani 1993). This is speculative, however, as immunohistochemistry and electron microscopy, as well as light microscopy, merely reflect differentiation, not histogenesis. It might thus be concluded that the histogenesis of PAS, as well as many other types of sarcomas, is poorly understood, and at the current state of knowledge, origin from a multipotent (undifferentiated) mesenchymal cell in the wall of the vessel is most likely (Nonomura et al. 1988; McGlennen et al. 1989; Burke and Virmani 1993). In most instances only autopsy material, variably preserved, has been available for the histopathological diagnosis which explains the designation of many cases as "undifferentiated sarcomas". This, of course may be true, like our cases 2 and 3; the tumours may be of the relatively non-specific fibro- myofibroblastic type that only express vimentin and smooth muscle specific actin. This finding may, in fact, lend support to the theory of an origin from a multipotent (undifferentiated) cell in the wall of the vessel.

References

Allen PW (1990) Primary leiomyosarcoma of the truncus-pulmonalis: report of a case with typical features and unusual metastases – letter to the case. Pathol Res Pract 186:300–301

Baker PB, Goodwin RA (1985) Pulmonary artery sarcomas: a review and report of a case. Arch Pathol Lab Med 109:35–39

Bleisch VR, Kraus FT (1980) Polypoid sarcoma of the pulmonary trunk: analysis of the literature and report of a case with leptomeric organelles and ultrastructural features of rhabdomyosarcoma. Cancer 46:314–324

Bolen JW, Hammar SP, McNutt MA (1986) Reactive and neoplastic serosal tissue: a light-microscopic, ultrastructural and immunohistochemical study. Am J Surg Pathol 10:34–47

Burke AP, Virmani R (1993) Sarcomas of the great vessels: a clinicopathologic study. Cancer 71:1761–1773

Dossche K, Wellens F, Goldstein JP, Deferm H, Bekaert Y (1992) Pulmonary homograft replacement for primary leiomyosarcoma of the pulmonary artery. J Thorac Cardiovasc Surg 104:844–846

Erlandsson RA (1981) Diagnostic transmission electron microscopy of human tumors. Masson, New York

Eyden BP, Banerjee SS, Harris M, Mene A (1991) A study of spindle cell sarcomas showing myofibroblastic differentiation. Ultrastruct Pathol 15:365–376

Fisher C (1990) The value of electronmicroscopy and immunohistochemistry in the diagnosis of soft tissue sarcomas: a study of 200 cases. Histopathology 16:441–454

Ghadialhy FN (1988) Ultrastructural pathology of the cell and matrix, 3rd edn. Butterworths, London

Head HD, Flam MS, John MJ, Lipnik SS, Slater DL, Stewart RD (1992) Long-term palliation of pulmonary artery sarcoma by radical excision and adjuvant therapy. Ann Thorac Surg 53:332–334

Hiroshima K, Uruma T, Ishibashi M, Ohwada H, Hayashi Y (1992) A case of primary sarcoma of the pulmonary artery. Acta Pathol Jpn 42:755–759

Johansson L (1989) Histogenesis of cardiac myxomas: an immunohistochemical study of 19 cases, including one with glandular structures and review of the literature. Arch Pathol Lab Med 113:735–741

Johansson L, Kugelberg J, Thulin L (1989) Myxofibrosarcoma in the left atrium developed from an endocardial myxoma with chondroid differentiation: a clinico-pathological report. APMIS 97:833–38

Kaiser LR, Urmacher C (1990) Primary sarcoma of the superior pulmonary vein. Cancer 66:789–795

Lundgren L (1993) Fibroblastic, myofibroblastic and myogenic spindle-cell lesions of a reactive and neoplastic nature. Thesis, Gothenburg

Mackay B, Floyd C, Ordoñez NG (1987) Ultrastructural observations on smooth muscle tumors. Ultrastruct Pathol 11:593–607

McConnell TH (1970) Bony and cartilaginous tumors of the heart and great vessels: report of an osteosarcoma of the pulmonary artery. Cancer 25:611–617

McGlennen RC, Manivel JC, Stanley SJ, Slater DL, Wick MR, Dehner LP (1989) Pulmonary artery sarcoma: a clinicopathologic, ultrastructural and immunochemical study of four cases. Mod Pathol 2:486–494

Nerlich A, Permanetter W, Ludwig B, Remberger K (1990) Primary leiomyosarcoma of the truncus-pulmonalis – report of a case with typical features and unusual metastases. Pathol Res Pract 186:296–299

Nonomura A, Kurumaya H, Kono N, Nakanuma Y, Ohta G, Terehata S, Matsubara F, Matsuda T, Asaka T, Nihsino T (1988) Primary pulmonary artery sarcoma: report of two cases studied by immunohistochemistry and electron microscopy, and review of 110 cases reported in the literature. Acta Pathol Jpn 38:883–896

Okada K, Okada M, Yamamoto S, Mukai T, Tsukube T, Matsuda H, Okada M (1993) Successful resection of a recurrent leiomyosarcoma of the pulmonary trunk. Ann Thorac Surg 55:1009–1012

Paulsen SM, Egeblad K (1983) Sarcoma of the pulmonary artery: a light and electron microscopy study. J Submicrosc Cytol 15:811–821

Ramp U, Gerharz CD, Iversen S, Schweden F, Steppling H, Gabbert HE (1992) Sarcoma of the pulmonary artery – report of 2 cases and a review of the literature. J Cancer Res Clin Oncol 118:551–556

Rangdaeng S, Truong LD (1991) Comparative immunohistochemical staining for desmin and muscle-specific actin: a study of 576 cases. Am J Clin Pathol 96:32–45

Shah IA, Kurtz SM, Simonsen RL (1991) Radiation-induced malignant fibrous histiocytoma of the pulmonary artery. Arch Pathol Lab Med 115:921–925

Smith WS, Lesar MS, Travis WD et al. (1989) MR and CT findings in pulmonary artery sarcoma. J Comput Assist Tomogr 13:906–909

Note added in proof

Since the preparation of this paper another case report has appeared (Nakazawa K, Itoh N, Shigematsu H, Kanbayasi T (1993) An Autopsy Case of Pulmonary Artery Leiomyosarcoma. Acta Pathologica Japonica 43:76–81). The authors present a case of high grade sarcoma positive for vimentin and alpha-smooth muscle actin, but not for desmin. On autopsy material an electron microscopical examination has been carried out which shows some cells with features of smooth muscle cells. The material is not very well preserved and the diagnosis is based on the discontinuous basement membrane along some cells. According to our opinion the cells may also be interpreted as myofibroblasts, then the diagnosis would be malignant fibrous histiocytoma.