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Teratoma of the heart

A case report and review of the literature

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Summary. The clinical manifestations and pathological features of an extremely rare cardiac tumour, a mature (benign) teratoma, in a 6 year-old girl are described. These are compared with those reported in the literature.

Key words: Cardiac tumour – Teratoma of the heart

Introduction

Primary tumours of the heart, in general, are considered rare, and teratomas in that organ are rather uncommon (Biancaniello et al. (1982); Larrieu et al. (1982); McAllister and Fenoglio (1978)). We report a case of a mature (benign) cardiac teratoma and review the literature on the subject.

Clinical history

This 6 year-old Tunisian girl (S.B.A.) was born on September 10th, 1975, after a normal pregnancy. Three children in the family are in good health. She was first seen in October 1980 by her physician in Tunisia, when she presented signs of an infection and heart failure, manifested by tachycardia and tachypnea. There was cardiomegaly and hepatomegaly with hepatojugular reflux as well as oedema of the lower extremities. A diagnosis of purulent pericarditis was made and antibiotics (Penicillin and Gentamycin) were administered for 30 days with a favourable outcome. Repeated echocardiographic examinations, during this period, revealed a cystic mass in the right ventricle, fixed to the thickened intraventricular septum. On auscultation, there was a 3/10 systolic murmur situated in the pulmonary area and the electrocardiogram showed a sinus rhythm at 110/min with complete right bundle branch block.

Catheterisation revealed low pulmonary arterial and capillary pressures with a slight gradient of 10 mm Hg, and confirmed the presence of a mass in the right ventricular cavity, attached to the upper part of the septum. The diagnosis of an echinococcus cyst of the right upper portion of the ventricular septum was proposed.

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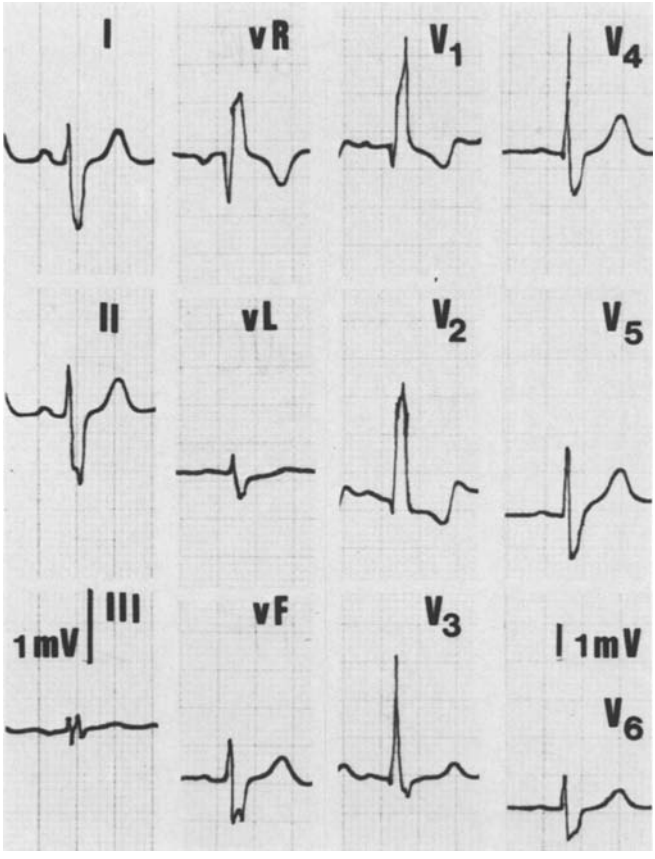


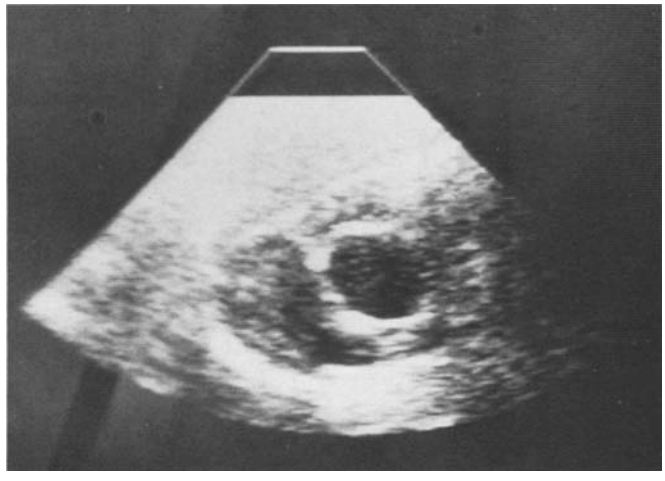
Fig. 1. Twelve lead electrocardiogram showing complete right bundle branch block and some degree of right ventricular hypertrophy

The girl was first seen at the Paediatric Department, Geneva, on November 18th, 1981. She weighed 19.2 kg, her heigth was 112.5 cm. Her blood pressure was 110/65 mm Hg and her pulse was regular at 104/min. There was no cyanosis, no chest pains, nor dyspnoea on effort. The arteries of the lower limbs were palpable.

On auscultation, there was a 4/6 systolic murmur with its maximum in the third intercostal space radiating slightly to the back with a widely split second sound. A chest roentgenogram showed normal lung fields but cardiomegaly, predominantly of the right ventricle. The electrocardiogram confirmed right ventricular hypertrophy with right atrial dilatation (Fig. 1) and complete right bundle branch block. Ultrasonography showed a multicystic mass in the right ventricle fixed to the upper portion of the interventricular septum and extending up to the atrio-ventricular septum (Fig. 2a and b). The CT-scan confirmed the presence of a multicystic mass, with some calcifications, fixed to the interventricular septum and protruding into the right ventricular cavity with partial obliteration of the cavity and infundibulum.

Cardiac catheterisation disclosed a patent foramen ovale without a shunt. The pressure in the pulmonary trunk was low with a gradiant between the pulmonary trunk and the right ventricle of 20 mm Hg to 25 mm Hg. On angiography the right ventricle was occupied by a large tumour fixed to the interventricular septum and with each contraction this came into contact with the opposing free right ventricular wall. The coronary arteries and their branches were normal except for the septal branches which were deviated in the area of the tumour.

a



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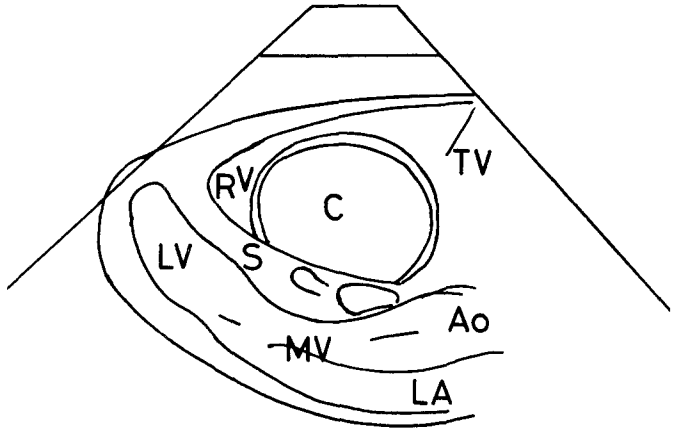

Fig. 2a, b. Cross-sectional echocardiogram and corresponding schematic drawing, clearly demonstrating a cystic tumor (*C*) in the right ventricle (*RV*). *LV*=left ventricle, *TV*=tricuspid valve, *MV*=mitral valve, *A*=aorta, *LA*=left atrium

There was no visualisation of vessels within the tumour. The sinus nodal artery took its origin from the main left coronary artery.

The white blood cell count was 15.750/mm^3 with a differential of 21.5% lymphocytes, 67% polymorphonuclears, 3.5% non segmented polymorphs, 0.5% eosinophils, 2% basophils and 5.5% monocytes. Haematocrit, haemoglobin, and platelet counts were normal. The blood group was 0 positive. ASLO measured 1,280 U and serology for echinococcus (ELSA) was neagtive. Urine and stool examinations were unrevealing. Sedimentation rate was 25/57 mm.

A probable diagnosis of hydatic cyst of the interventricular septum was made.

The girl was discharged after 2 days and readmitted one month later to the Paediatric Surgical Department for cardiac surgery. At operation, a whitish cystic mass situated in the upper part of the right side of the inter-ventricular septum and protruding into the infundibulum was observed. Two of the cavities were punctured. They contained a thick, brownish, gelatinous fluid. A third cyst contained a clear fluid. There were neither scoleces nor bacteria in the material. In the belief that this was a hydatid cyst, hypertonic saline was injected into the cavities. Total excision of the mass was then undertaken leaving a cavity, but the

septal papillary muscle was fixed to the septum. The patient developed severe pulmonary oedema which became haemorrhagic and she died 3 h after the operation.

Pathology

Post-mortem (A 912/81) was performed 18 h after death. The body weighed 17.200 kg for a height of 112.5 cm, and was in relatively good nutritional condition. The main pathological findings were confined to the heart and lungs.

Heart The pericardial sack was open, containing numerous large blood clots mixed with about 50 ml of haemorrhagic fluid. The heart weighed 190 g, was firm in consistancy and presented a suture line of 4.5 cm on the anterior surface of the right ventricule. The latter, dilated and completely necrosed, was purple in colour. The right atrium was dilated and partially necrosed. The septal leaflet of the tricuspid valve was resected and a cavity of 2.8 cm in diameter, representing the site of the tumour, occupied the inlet and trabecular portions of the interventricular septum including the medial papillary muscle as well as the posterior limb and a portion of the body of the trabecula septomarginalis. The left ventricle was moderately hypertrophied but its cavity, like that of the left atrium, was not dilated. The aorta, pulmonary trunk and their corresponding valves were normal, as well as the remaining portions of the tricuspid valve. The coronary arteries took their origin normally and were without lesions.

Lungs. Each pleural cavity contained approximately 250 ml of haemorrhagic fluid. Both lungs were atelectactic, the right weighing 310 g, the left 285 g. They showed haemorrhagic consolidation.

Tumor (T 18507/81). Gross pathology: The resected, lobulated, cardiac mass measuring $4.2 \times 3.5 \times 2.5$ cm was recovered by a thick, whitish capsule. An X-ray showed a central calcified nodule. The cut surface presented several cystic cavities varying between 2 mm and 3 mm to about 3 cm in diameter and containing either a thick gelatinous, light-brown fluid or a clear liquid. The wall was thin in places, but measured up to 1.2 cm in others with some hard areas.

Histology

The tumour was fixed in 10% neutral buffered formalin. Whole transverse and other sections were taken for histology, embedded in paraffin wax, stained with Haematoxylin-Eosin, Verlhoff-van Gieson, Masson's trichrome and PAS stains.

The largest cystic cavity containing eosinophilic material was partially lined by a pseudostratified columnar or cylindrical epithelium. Part of the wall of the cavity was occupied by lymphoid tissue which was covered

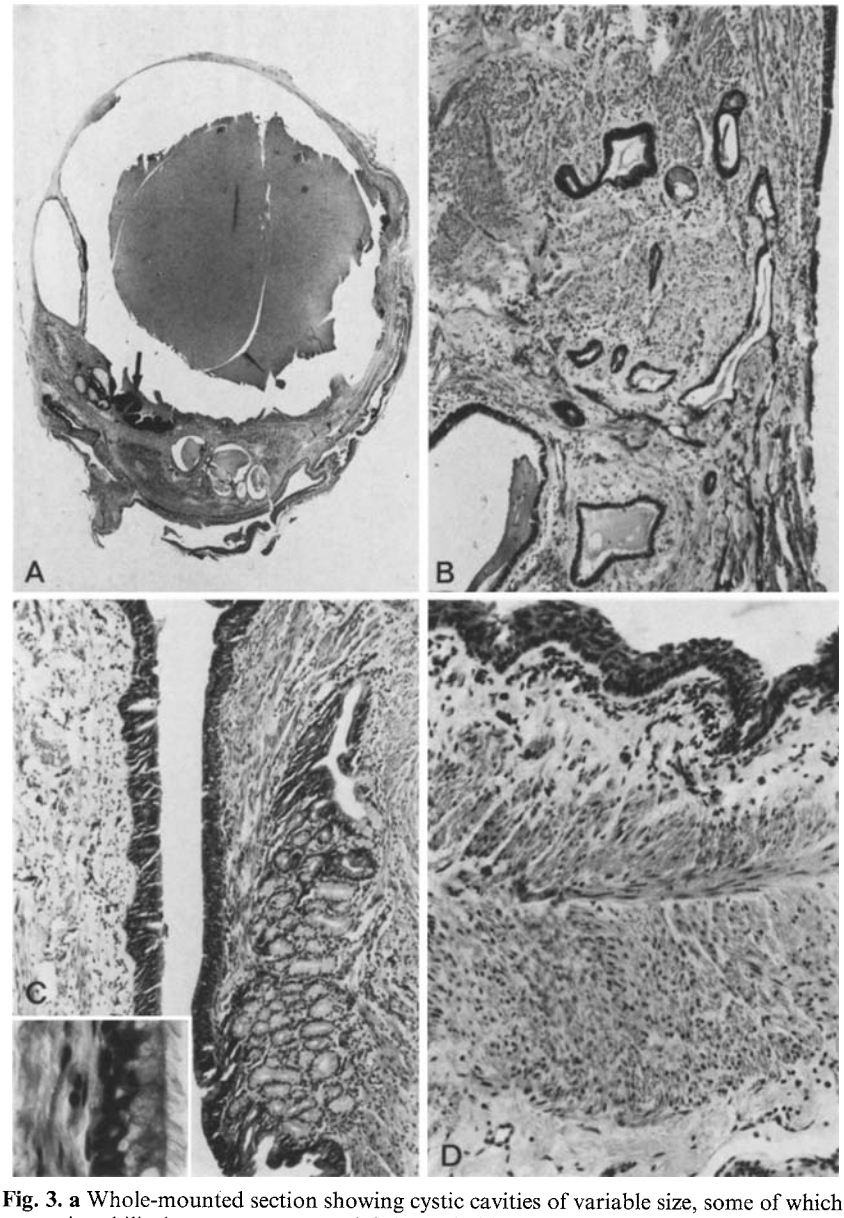

Fig. 3. a Whole-mounted section showing cystic cavities of variable size, some of which contain an eosinophilic homogenous material. Lymphoid tonsillarlike areas (*arrowed*) border some cysts. **b** In a dense connective tissue stroma containing smooth muscle bundles are numerous small cysts lined by a tall columnar epithelium (H.E. × 30). **c** In the neighbourhood of some of the cystic cavities are groups of mucus glands which sometimes open into the lumen. The pseudo-stratified epithelium is ciliated (*inset*) (H.E. × 60). **d** Occasionally, well-developed smooth muscle bundles completely surround the cystic cavities giving them an organoid appearance (H.E. × 120)

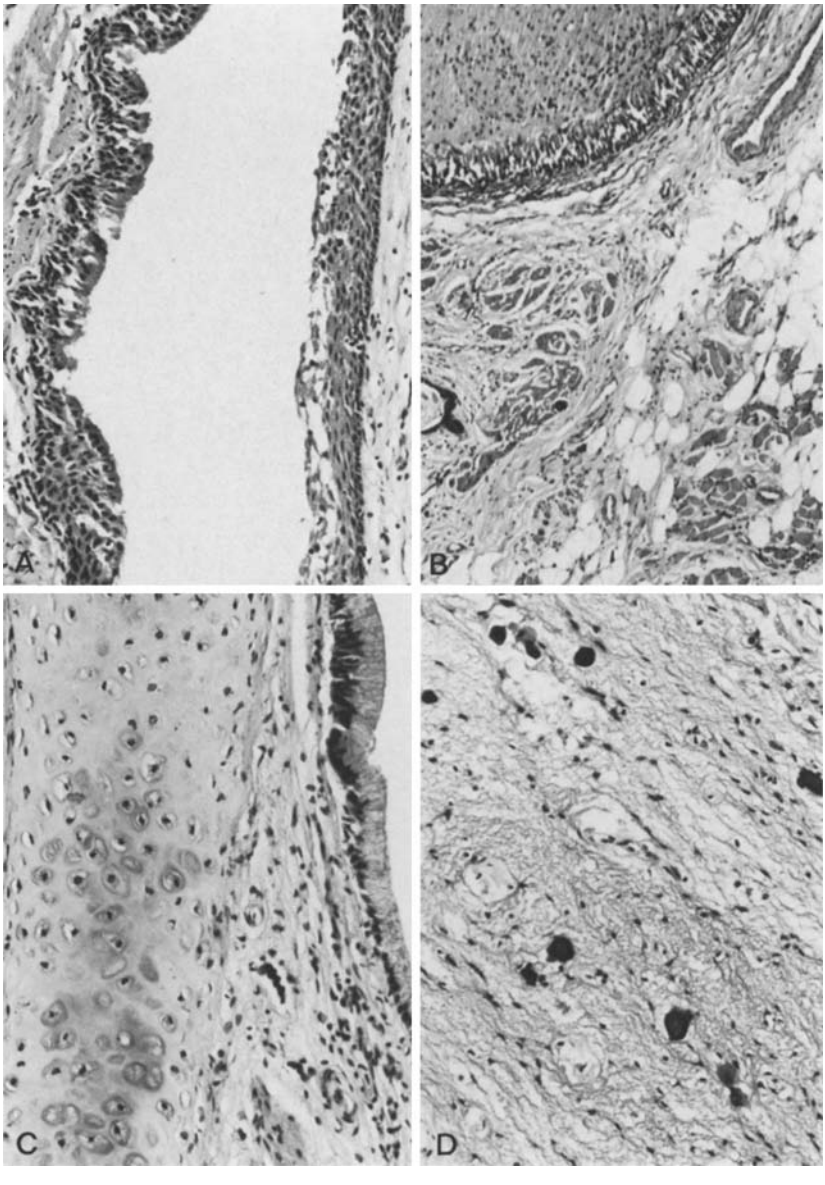

Fig. 4. **a** Some cysts are lined partly by squamous epithelium (H.E. ×60). **b** Nests of striated muscle bundles together with some fatty tissue are observed in the vicinity of glandular structures in association with smooth muscle bundles (H.E. ×30). **c** Large hyalin cartilage plates border some of the cavities lined by tall ciliated columnar epithelium (H.E. ×60). **d** Neuroglial tissue containing calcified masses are randomly scattered throughout the stroma (H.E. ×60)

by cylindrical or squamous epithelium, the whole imitating tonsillar tissue (Fig. 3a). In the adjacent stroma there were numerous cysts of variable size (Fig. 3b), which were lined for the most part either by a cylindrical, ciliated epithelium or a pseudostratified epithelium or a combination of both (Fig. 3c). Mucus glandular structures opened in some of these cysts which were surrounded by layers of smooth muscle bundles of variable thickness (Fig. 3d). A few glands were lined by squamous epithelium (Fig. 4a). There were also mixed glandular structures resembling salivary glands. Between these glands there were varying amounts of smooth muscle bundles, striated muscle fascicles, cartilage plates but no bone formation (Fig. 4b and c). Large areas of neuroglial tissue containing calcified masses were scattered throughout the tumour (Fig. 4d). Fat tissue was scanty. Blood vessels were plentiful and few nerve sheaths were present containing, on occasion, isolated groups of ganglion cells. No immature structures were observed in the many sections examined.

At one of the margins, the fibrous tissue continued into the adjoining myocardium which showed atrophic changes in the myofibrils. The general histological appearances were consistant with those of a mature (benign) teratoma with all three germ layers present.

Discussion

Primary tumours of the heart are considered extremely rare and it has been estimated that their overall incidence varies between 0.0017% and 0.28% (McAllister and Fenoglio (1978)). In infancy and childhood primary cardiac tumours are exceptional and up to recently were considered incidental post-mortem findings (Arciniegas et al. (1980a); Schmaltz and Apitz (1981)). With the advent of modern techniques (invasive and non-invasive) it is now possible to diagnose such lesions during life, thus offering the possibility of surgical treatment with a favourable outcome in the majority of cases (Biancaniello et al. (1982); Chadraratina et al. (1977); Larrieu et al. (1982)).

Although teratomas and cysts of the anterior mediastinum are not altogether uncommon, forming about 20% of tumours and cysts of the region (Carter et al. (1982); Wychulis et al. (1971)), they are relatively rare in the heart and pericardium. Fifty-seven cases of intrapericardial teratomas have been described in the literature (Arciniegas et al. (1980b); McAllister and Fenoglio (1978)), whereas we were able to find only 9 documented cases of teratomas or possible teratomas localized in the heart. The pertinent clinical and pathological findings of these cases are summarized in the Table 1. The tumours were accepted as genuine teratomas only when they presented elements of the three (ectoderm, mesoderm endoderm) germ layers in histological sections; thus all epithelial and/or bronchogenic cysts of the heart were excluded. Of the 10 cases, including the one described here, all except two patients, a 53 year-old female and a 56 year-old male, were in the paediatric age group. There were 7 females and 3 males (2.3:1). It is interesting to note that of the 14 intrapericardic teratomas described

Table 1

Case no	Author	Sex	AGE	Localisation	Size	Histology	Remarks
1	De Châtel (1933)	F	New-born	Right atrium along interauricular septum in continuity with the tricuspid valve	Lentille (pea)	Cystic cavities lined by flat or pluristratified epithelium with areas of cylindric epithelium. Presence of sudoriparous glands and blood vessels	Associated meningoencephalocele. Incomplete fermature of the nasal septum
2	Anderson and Dmytryk (1946)	F	53 years	Right atrium extending to tricuspid valve	8 × 5 cm	Cystic spaces up to 1–5 cm in diameter. Loose myxomatous stroma with sarcomatous appearance. Mitotic figures. Invasion of myocardium and epicardium. Areas of bone formation. Gland-like structures lined by columnar or secretory cells with papillary structures. Ciliated epithelium	Malignant teratoma with local invasion
3	Leighton et al. (1950)	F	6 months	Right atrium at the base of the medial tricuspid leaflet	2.5 × 2 × 1.5 cm	Multilocular cysts up to 1.5 cm in diameter, lined by stratified squamous epithelium. Undifferentiated mesenchymatous stroma with fibroblast and foreign body giant cells	Large interventricular communication
4	Solomon (1951)	F	2 years	Interauricular septum protruding into right auricle and invading myocardium	Not given	Lobulated mass with necrotic areas and cystic cavities. Respiratory elements, some squamous epithelium and sebacous glands	Patent foramen ovale. Malignant teratoma with metastases to lung, skull and brain
5	Williams (1961)	M	20 days	Right ventricle filled with tumour extending into pulmonary outflow track	2 cm in diameter	Multicystic cavities with derivatives of all three germ layers	Patent foramen ovale

6	Arshadi and Watson (1966)	F	4 years	Origin from right ventricular septum projecting into and filling the right ventricular cavity	9 × 8 × 8 cm	Two cell types a) cyto- and syncytiotrophoblastic areas of choriocarcinoma with a female chromatin dot in the nucleus. b) abundant small basophilic immature-appearing epithelial cells. Both types were embedded in a fibrous stroma	Malignant teratoma (teratocarcinoma) with lung metastases
7	Gerbode et al. (1967)	F	8 years	Anterior wall of right ventricle involving papillary muscle and base of anterior tricuspid leaflet	Not given	Right ventricular tumour attached to the anterior wall, papillary muscles and anterior tricuspid leaflet	Resected
8	Van der Hauwaert (1971)	M	15 days	Right atrial and ventricular cavities extending through tricuspid orifice with insertion in the inter-atrial an inter-ventricular septum	Huge multi-locular cystic mass	Derivatives of all three primitive germ layers	
9	Cabanas and Moore (1973)	M	56 years	Arose in right ventricle from a broad base on the inter-ventricular septum and adjacent myocardium of the anterior wall and extending through the pulmonary valve producing partial obstruction	7 × 3 cm	Composed predominantly of sheets and cords of poorly differentiated epithelial cells forming glandlike structures in an acellular fibrocollagenous myxomatous stroma. Nests of stratified squamous epithelium in highly cellular areas with mitotic figures, cartilage islands and bone formations	Malignant teratoma with metastases to the thoracic and lumbar vertebrae
10	Present case	F	6 years	Upper portion of right ventricular septum occupying the inlet and trabecular portions of the inter-ventricular septum	4.2 × 3.5 × 2.5 cm	Multilocular cysts up to 3 cm in diameter. Derivatives of all three germ layers	Patient died after resection of tumour

by McAllister and Fenoglio (1978), there was only one male, while Arciniegas et al. (1980b) in their review of the literature on the subject did not register any significant sex preponderance.

All the teratomas of the heart so far described have been located in the right heart; 4 in the right atrium located mainly in the interauricular septum (cases 1, 2, 3 and 4), 5 in the right ventricle (cases 5, 6, 7, 9 and 10) and one involving both the interauricular and interventricular septa (case 8). Although the dimensions of the tumours were not always given, there was great variability among those recorded, varying between 5 mm and 9 cm or more in greatest diameter. The tumours were often lobulated containing many multilocular cystic cavities of variable diameter. The cystic contents were recorded either as clear, yellow or brownish, and in the same tumour the colour varied from one cavity to the other as in the case described here.

Histologically the cysts were lined by various epithelial structures, squamous in places, flat cuboidal in others or pluristratified. Cylindrical ciliated epithelium was also a common feature in some instances (Anderson and Dmytryk (1946); Solomon (1951); Williams (1961)). The stroma was sometimes reported as myxomatous but more often it was composed of loose or dense fibrous tissue with some muscle bundles (smooth and/or striated), blood vessels and nerve fibres. Sometimes smooth muscle bundles formed collars around glandular structures and together they resembled respiratory elements as in this case. Cartilage was often noticed in the vicinity of these structures (Cabanas and Moore (1973)).

With the exception of 4 cases (cases 2, 4, 6 and 9), the remaining 6 were benign tumours with mature elements. In case No. 2 most of the tissue was myxomatous with few cells in an abundant loose, pale staining stroma (Anderson and Dmytryk (1946)) or taking on a loose fibrillar structure around numerous capillaries. There were, however, highly cellular areas having a sarcomatous appearance with infiltration of the adjacent myocardium and reaching the epicardium in places, indicating local invasion and thus the malignant nature of the tumour. In case No. 4 (Solomon (1951)) metastasis to the right supraorbital region was the presenting feature and at post-mortem the tumour had not only invaded the interauricular septum but had produced metastases to the right frontal bone with invasion of the cranial cavity and overlying dura. Case No. 6 presented in areas as a choriocarcinoma with cyto- and syncytiotrophoblastic elements associated with immature appearing epithelial cells. There were metastases to the lung (Arshadi and Watson (1966)). Case No. 9 (Cabanas and Moore (1973)) was a poorly differentiated tumour in a myxomatous stroma englobing nests of squamous epithelium. Mitotic figures were numerous but some areas contained cartilage plates with bone formation.

Cardiac malformations or other abnormalities have been described in association with cardiac teratomas. A patent foramen ovale has been reported by Solomon (1951) and Williams (1961) while Leighton et al. (1950) observed a large interventricular septal defect. De Châtel's (1933) case had a meningo-encephalocele assoicated with an incomplete closure of the nasal septum.

On morphological grounds teratomas of the heart must be distinguished from other benign or malignant tumours (Heath (1968); Larrieu et al. (1982); Silverman (1980)). In general, one must consider the epithelial cysts of the myocardium which are very often situated in the thickness of the left ventricular wall or atrioventricular node (Marshall (1957); Travers (1981)), as well as other more common tumours of the heart (myxoma, rhabdomyoma, fibroma, lipoma and the sarcomas (rhabdomyosarcoma, angiosarcoma, fibrosarcoma)), (Biancaniello et al. (1982); Ilbawi et al. (1982); Larrieu et al. (1982); Routon et al. (1979)). Of special interest are the coelotheliomas or dysplastic tissues encountered in the atrioventricular node and surrounding tissues due to their possible embryological origin. These tumours create early bundle branch block (Doerr (1982); Mahaim (1945)). The clinical signs are usually non specific and depend largely on the location and size of the tumour (Arciniegas et al. (1980a); Biancaniello et al. (1982); Wold and Lie (1980)). Roentgenography is usually of little assistance in arriving at a diagnosis, whereas angiography and especially contrast angiography, gated cardiac pool radionuclide imaging and computerized tomographic scan can be of considerable help in defining the structures, their precise location and size (Arciniegas et al. (1980a); Biancaniello et al. (1982)). Two-dimentional echocardiography has now made it possible to arrive at a diagnosis with great accuracy without the use of invasive techniques (Chandraratua et al. (1977); Oliver et al. (1982)) Surgery is the treatment of choice when the diagnosis is made during life. The results evidently depend on the size, localization and whether the tumour is benign or malignant (Arciniegas et al. (1980a); Attar et al. (1980; Biancaniello et al. (1982); Schmaltz and Apitz (1981); Ursell et al. (1982)). Very few cases of cardiac teratomas have so far been operated on and therefore one cannot arrive at a prognosis, but by analogy with the other cardiac tumours which have been operated upon, the results should be favourable.

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