

LETTER TO THE EDITOR

Sir,

Recently Leif Johansson and Brigitta Carlen [1] reported in this Journal on four sarcomas of the pulmonary artery and discussed their frequency, fine structure, histogenesis and classification. It is by chance that we, at the Institute of Pathology of the University of Heidelberg, were in possession of a very impressive collection specimen of such a peculiar tumour which we called “embolus-like sarcoma of the pulmonary artery (emboliformes Sarkom der Pulmonalis)”. The late Professor Curt Froboese presented and discussed this case at the 90th Meeting of the German Scientists and Physicians in Hamburg, September 21, 1928. He also described it in detail in a paper published in the *Zentralblatt für Pathologie* [2]. At that time Curt Froboese was Privatdozent at the Institute of Pathology in Heidelberg, which was then under the directorship of Paul Ernst. The unique exhibition specimen of this tumour was unfortunately lost when the Institute moved into a new building in 1966. However, the specimen was so impressive and instructive that, in 1979, I was able to make a *prima vista* diagnosis of another case of sarcoma of the pulmonary artery. This case was published by Henrichs et al. in this Journal [3].

In connection with these cases a comment made by R. Rössle in the discussion at the Naturforscher Meeting in

Hamburg 1928 is of interest. He emphasized the relationship of Froboese's tumour with a leiomyosarcoma of the conus pulmonalis described by Eschbach [4]. Figure 1 is a reproduction of the gross pathology of the tumour published by Froboese; Fig. 2 shows the histological appearance of the tumour described by Eschbach. What I would like to point out here, is the possible relationship of these peculiar tumours with atherosclerotic lesions described by Benditt and Benditt (reviewed in 5). These authors discussed that certain types of atherosclerotic intima lesions – those with multifocal cellular proliferations which probably originate from the Langhans-Wissler-cells, are of monoclonal nature. Benditt saw in this lesion something like a benign neoplasm. I have much sympathy with this interpretation and believe that these intimal changes could initiate an autonomous cell proliferation. The article by Johansson and Carlen gives me the opportunity to stress the possible histogenetic connection between “Benditt's arteriosclerotic lesion” and

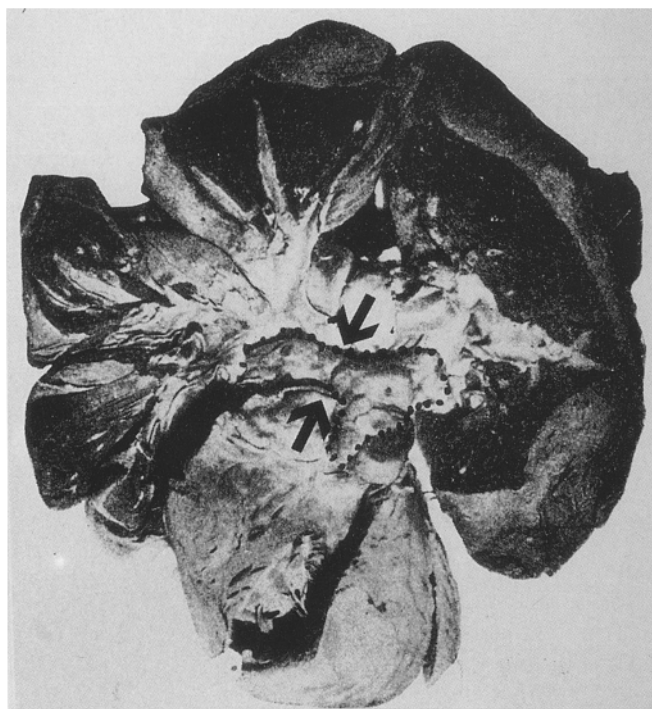


Fig. 1 Case described by Froboese [2]. 64-year-old woman with primary sarcoma of the intima of the pulmonary artery. Tumour size: 5×3×3 cm. The illustration shows the right heart with a tumour in the pulmonary trunk imitating a thrombembolus (arrows)

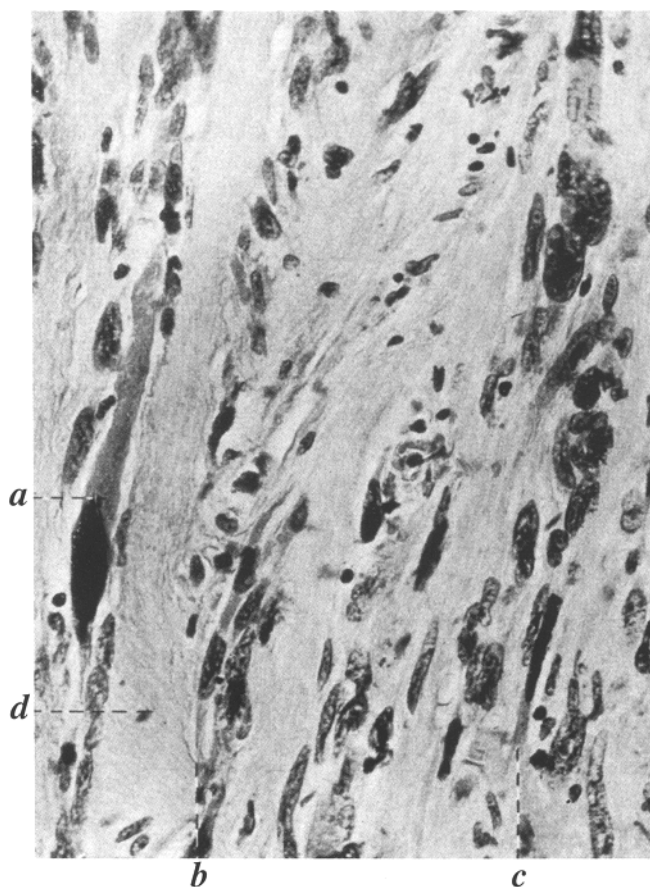


Fig. 2 Case described by Eschbach [4]. 58-year-old woman who had an occluding tumour in the pulmonary trunk. The illustration shows a polymorphocellular sarcoma reminiscent of a leiomyosarcoma

the rare sarcomatous neoplasia of the large arteries, a relationship which I discussed in more detail in *Documenta Angiologorum* [6].

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References

1. Johansson L, Carlen B (1994) *Virchows Arch* 424:217–224
2. Froboese C (1928) *Zbl Pathol* 44:148–153
3. Henrichs KJ, Wenisch HCJ, Hofmann W, Klein F (1979) *Virchows Arch [A]* 383:207–216
4. Eschbach H (1928) *Beitr Pathol Anat* 80:672–681
5. Doerr W (1978) *Virchows Arch [A]* 380:91–100
6. Doerr W (1990) *Documenta Angiologorum* Vol XXI:6–25

REPLY

Sir,
in this journal we recently reported the clinicopathological features of four cases of pulmonary artery sarcoma that appeared at our institution during a period of 30 years [3]. In a letter to the editor Professor Doerr relates two cases of his own experience, one described by Froboese in 1928 and termed “embolus like sarcoma of the pulmonary artery” and the other published by Henrich et al. [2] Professor Doerr also discuss the possible histogenetic relationship between sarcomas of the pulmonary artery and certain arteriosclerotic lesions, namely multifocal cellular proliferations.

If this relationship is a real one sarcomas should appear relatively often in the large systemic arteries as arteriosclerosis is a frequent disease in these vessels. However, sarcomas of the large vessels are exceedingly rare, perhaps least so in vena cava inferior and arteria pulmonalis, and most so in the large systemic arteries [4]. There is no evidence that pulmonary artery sarcoma appears more frequently in patients with pulmonary hypertension, the only disease in which pulmonary artery arteriosclerosis is found with some regularity. It is possible to have much sympathy with the hypothesis that “blastomatous cellular proliferation” be regarded as a benign neoplasm and a kind of a missing link between arteriosclero-

sis and malignant neoplasms (sarcomas) of the great vessels [1] but I do not think that it can fully explain the exceedingly rare appearance of malignant neoplasms in the great vessels. This becomes even more obvious when considering the divergent histopathological appearance of these neoplasms; either this is regarded as differentiation or histogenesis [3].

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References

1. Doerr W (1990) *Documenta Angiologorum* XXI
2. Henrich KJ, Wenisch HJC, Hofman W, Klein F (1979) Leiomyosarcoma of the pulmonary artery: A light and electronmicroscopical study. *Virchows Arch [A]* 383:207–216
3. Johansson L, Carlén B (1994) Sarcoma of the pulmonary trunk: Report of four cases with electron microscopic and immunohistochemical examinations, and review of the literature. *Virchows Archiv* 424:217–224
4. Kaiser LR, Urmacher C (1990) Primary sarcoma of the superior pulmonary vein. *Cancer* 66:789–795