

CASE REPORTS

BALL-VALVE MITRAL OBSTRUCTION BY A SARCOMA OF A PULMONARY VEIN

BY

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Primary tumours of major blood vessels are rare. Giaccai (1949), in his review of reported cases found 9 benign tumours, one of which was an enchondroma of a superficial vein of the forearm, and 17 malignant tumours, four of which were sarcomata of the pulmonary artery, and one (Ausbüttel, 1939) from the right pulmonary vein. This latter had the structure of a pleomorphic-celled sarcoma, proliferated as far as the left atrium, and gave rise to an implantation metastasis on the anterior mitral leaf. Ravid and Sachs (1943), reported a primary myxosarcoma of the left auricle and pulmonary vein associated with multiple tumours of various types elsewhere in the body. We report here a tumour of the right superior pulmonary vein which grew into the left atrium and occluded the mitral opening.

Case Report

W. H., a 47-year-old farmer, was quite fit until October, 1954, when he had a febrile respiratory illness with hæmoptysis. There were no physical signs in the chest or signs of recent thrombophlebitis. X-rays showed a small hazy shadow in the right mid-zone. He recovered and returned to work but was readmitted with hæmoptysis the following month. The shadow was now resolving and physical and cardiographic examinations were negative. Subsequently he felt well apart from an ache in the right side of the chest.

Four months after his first illness he was readmitted to hospital with persistent hæmoptysis and extreme dyspnœa accompanying attacks of coughing. He was cyanosed and the physical and radiological signs were those of pulmonary œdema. His pulse was very rapid and small and he showed intermittent auricular fibrillation. The first sound at the mitral area had a slapping quality. He died within a few hours.

Necropsy Findings. There was some congestion of the bronchi and lower part of the trachea and both lungs were congested and œdematous. About 500 ml. of faintly brownish fluid were present in the left pleural cavity. The right pleural cavity was almost obliterated by old adhesions but nevertheless contained some excess fluid. The liver showed a slight nutmeg pattern. The spleen was a little enlarged. The major arteries had only a few small flecks of atheroma. All other organs except the heart were normal.

Heart. The left atrium was found to be filled by a tumour measuring 5.0 cm. in maximal diameter (Fig. 1). It had a slightly roughened surface and bulged into the mitral orifice. The pericardium, myocardium, endocardium, and valves were normal. Further dissection showed that the tumour arose from a small pedicle in the right superior pulmonary vein between 2 and 3 cm. away from the vein's junction with the left atrium. It almost occluded the lumen of the vein and in many places was adherent to the intima. This adherence was only slight and separation could easily be effected by gentle stripping except at two sites, one at the veno-atrial junction, and the other shortly after its origin, where the attachment was firm. In parts the tumour was fairly soft, but most of it was hard and could not be cut with a knife.

Microscopically it was found that the pedicle was composed of collagen, a few elastic fibres, and material that was metachromatic with toluidine blue and removable by hyaluronidase. Adjacent to its pedicle the growth consisted of well formed non-malignant osseous tissue but this rapidly gave place to an osteogenic sarcoma which constituted its remainder (Fig. 2). At the veno-atrial junction there was invasion of the intima of the vein and atrium but this did not penetrate beyond the elastic lamina. At the other site of firm attachment the growth penetrated through the vein wall and spread to a slight extent in the surrounding

lung tissue. A blood vessel supplying the tumour opened at the base of the pedicle and can be seen in Fig. 3. Elsewhere the blood supply seemed to be derived from vessels opening directly on the surface.

At the point of origin of the pedicle the pulmonary vein received a tributary which was not normal: this tributary was excessively convoluted and had marked fibroelastic thickening of its intima. Other blood vessels in the lungs seemed satisfactory except for a few on the right side which showed evidence of thrombosis and recanalization. Both lungs were congested.

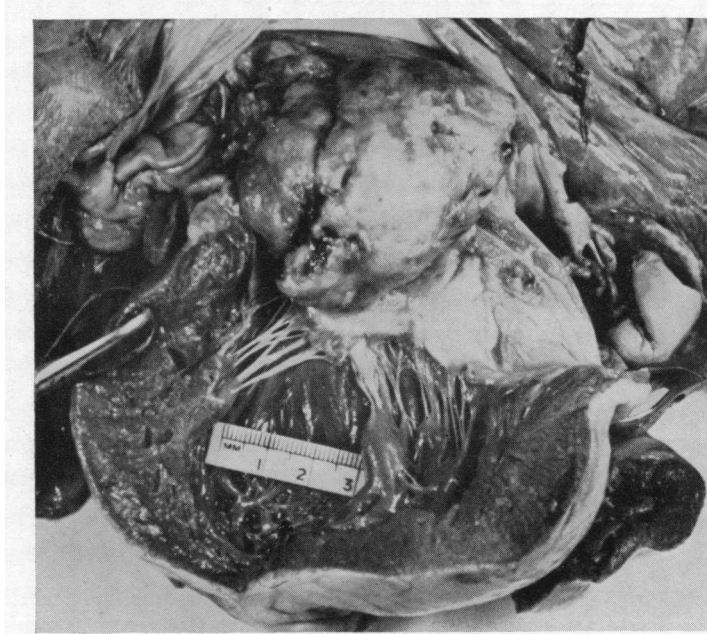


FIG. 1.—Interior of heart, showing polyp obstructing mitral orifice.

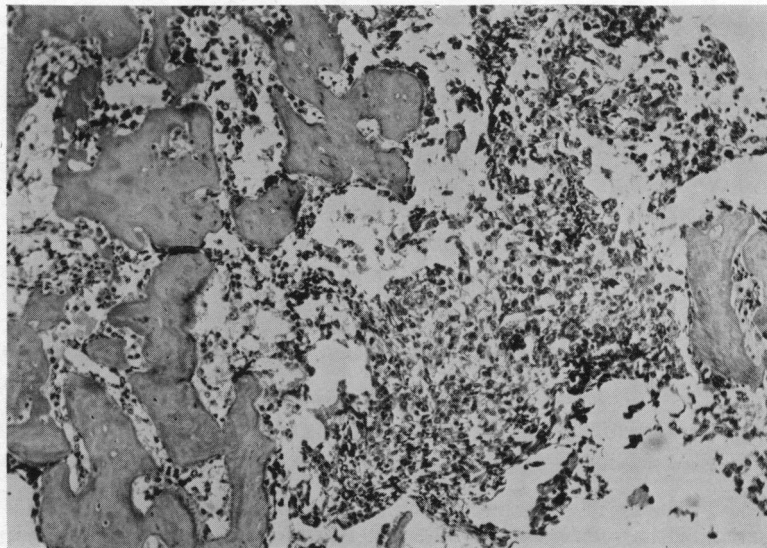


FIG. 2.—Osteosarcomatous tissue. Hæmatoxylin and eosin : $\times 85$.



FIG. 3.—View of pedicle, showing tortuous tributary at base. Elastic and van Gieson: $\times 3.2$.

Discussion

The osteogenic sarcomatous nature of the tumour was not realized at the time of the post-mortem examination, and an extensive examination of the bones was not carried out. Nothing suspicious was found in those bones examined, however, and the patient had no bone symptoms during life. The pulmonary vein is a highly unlikely site for a metastasis, and where there was slight penetration through the wall into the surrounding lung tissue, the appearances did not suggest that the vein had been invaded from without. The very structure of the tumour in fact suggests that it had originally been a benign polyp containing bone tissue which underwent malignant change.

Multiple ossifying cartilaginous nodules attached by fibrous pedicles within the left ventricles of rats were described by Farris *et al.* (1946). Bone marrow was found in some of them. The authors assumed that the changes were due to the metaplasia of fibroblasts in areas of degeneration or scarring from an unknown cause. It is possible that the lesions were congenital. Prichard (1951) suggested that myxomata of the left atrium arose from small malformations in the region of the fossa ovalis. These malformations consisted of excess fibrous tissue and in some cases minute areas where there were lacunæ of capillary size lined by plump endothelial cells. This suggestion has been supported by Raeburn (1952) who draws a comparison with epidermal moles. Myxomata sometimes contain bone. One of Prichard's cases had at its base large thick-walled blood vessels arising from deep within the atrial wall. Giacciai supported the theory of dysontogenetic neoplasia for some blood vessel tumours, particularly those arising from the pulmonary vessels, as they are in a zone where evolutive errors can occur frequently.

In our case the pedicle was situated where the pulmonary vein received an abnormal tributary, and we suggest that this patient had a small bone-containing hamartoma which eventually became malignant just as a skin nævus may become a malignant melanoma. The plasticity of mesenchymal tissue and its potency for aberrant differentiation is now generally acknowledged (Forbes, 1955).

Summary

A case is described of ball-valve obstruction of the mitral orifice by an osteogenic sarcoma of a pulmonary vein. The tumour was probably derived from a pre-existing hamartomatous polyp.

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References

- Ausbüttel, F. (1939). *Frankf. Zeitschr. Path.*, **53**, 303.
 Farris, E. J., Yeakel, E. H., and Seitner, M. M. (1946). *Amer. J. Path.*, **22**, 613.
 Forbes, G. B. (1955). *J. Path. Bact.*, **70**, 427.
 Giacciai, L. (1949). *Arch. de Vecchi anat. pat.*, **13**, 353.
 Prichard, R. W. (1951). *Arch. Path.*, Chicago, **51**, 98.
 Raeburn, C. (1952). *J. clin. Path.*, **5**, 339.
 Ravid, J. M., and Sachs, J. (1943). *Amer. Heart J.*, **26**, 385.