

Coexisting congenital primary superior vena caval aneurysm and rheumatic mitral stenosis

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Congenital primary superior vena caval aneurysm (CPSVCA) as classified by Abbott and Leigh¹ is extremely rare. Two types have been described—fusiform, which is more common, and saccular. It was first reported by Abbott in 1950.² To our knowledge, only 12 cases have been reported. In this article, we describe the fourth case of saccular CPSVCA and the first which coexisted with rheumatic mitral stenosis.

Case report

The patient (CFL) was a 48-year-old Chinese housewife. She was referred to us in January 1980 for treatment of a "mediastinal cyst." In November 1979, she was admitted to a general hospital with a week's history of dyspnoea on exertion. Her past medical history was non-contributory. On examination, she had the signs of congestive cardiac failure. Her chest radiograph on admission showed a large anterior mediastinal shadow extending into the right pleural cavity together with cardiomegaly, pulmonary congestion, and pleural effusion. A diagnosis of congestive cardiac failure and mediastinal tumour was made. At that time, although the cause of the cardiac failure was not identified, diuretic therapy was given with a dramatic response. Radiologically the pulmonary congestion and pleural effusion disappeared and there was a significant reduction in size of the mediastinal shadow. In view of this, the mediastinal mass was aspirated twice. On both occasions frank blood was obtained. An aortogram was then performed. The proximal aorta and its branches were found to be normal. In the meantime, facilities for ultrasound scanning became available at that hospital. Sonography showed a cystic lesion with well-defined borders. It was not connected to the heart or aorta. A diagnosis of "sanguineous mediastinal cyst" was made and she was referred to us for further management.

On her admission to our hospital, physical examination showed that the patient was also suffering from mitral stenosis, probably rheumatic in origin. This was confirmed by M-mode echocardiography and cardiac catheterisation. Her chest radiograph (fig 1) showed an oval anterior mediastinal shadow extending over the right hilar region, slight cardiomegaly, prominent pulmonary conus, and dilatation of the upper lobe veins.

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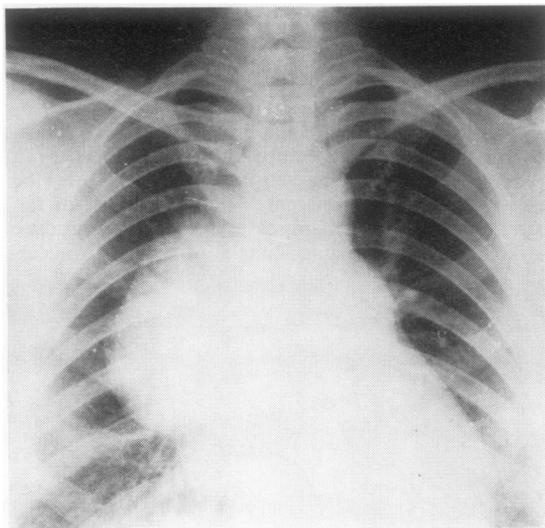


Fig. 1 Chest radiograph of the patient after she was treated with diuretic.

The patient was scheduled for excision of the "mediastinal cyst" and open mitral valvotomy. A median sternotomy was performed. A pear-shaped, bluish, softly cystic mass, 10 × 4 cm was found on the right side of the mediastinum. It lay loosely on the pericardium and could be readily separated from the right mediastinal pleura. On lifting up the fundus of the cystic mass, it exhibited characteristic systemic venous pulsations. Needle aspiration yielded venous blood. Further dissection revealed a 1 cm wide and 1.5 cm long stalk at its point of origin immediately above the junction of the azygos vein and the superior vena cava. The stalk was divided and the venous aneurysm was excised in toto. Open mitral valvotomy was performed under standard cardiopulmonary bypass. The patient's postoperative course was unremarkable, and follow-up at seven months was completely satisfactory.

Pathological examination of the resected specimen revealed an oblong sac measuring 6 cm longitudinally and 3 cm transversely. The outer wall of the sac consisted of wrinkled fibrous tissue, glistening in areas. The inner wall was generally wrinkled and trabeculated.

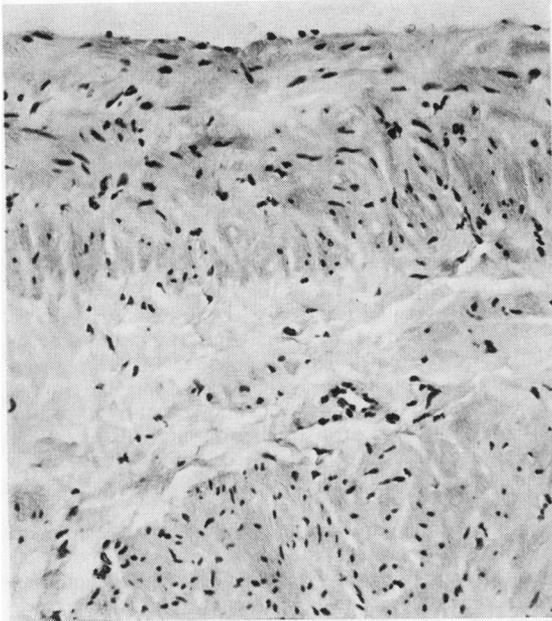


Fig 2 Microscopic appearance of the wall of the aneurysm. Original magnification H and $E \times 250$.

There was an indurated plaque 8×6 mm at the fundus of the sac. Histologically, the wall of the sac resembled that of a large vein in muscle arrangement and elastic fibres. There was an intima of well-defined endothelium and connective tissue, a media of inner circular and longitudinal muscle, as well as an adventitia of outer longitudinal muscle (fig 2). The muscle layers were much thicker than those found in superior vena caval samples of matching age and sex. The plaque at the fundus of the sac was composed of layers of fibrous tissue with fibrin layers at the centre. This probably represents a slow thrombotic process.

Discussion

Superior vena caval aneurysm is classified as congenital and primary when no other aetiological factor is recognised. The microscopic appearance of this type of venous aneurysm has only been described on three previous

occasions.³⁻⁵ The wall of the aneurysm was mainly fibrous and lined with endothelial cells. Apart from our case, smooth muscle has been found in only one other case.⁴

The clinical features, diagnosis, and management of CPSVCA have been well described.^{4, 5} The present case is interesting because it coexisted with rheumatic mitral stenosis. As a result, it disclosed the appearance and behaviour of a CPSVCA in the presence of congestive cardiac failure.

Most authors are of the opinion that CPSVCA should be managed conservatively, because they do not enlarge, produce pressure symptoms or rupture, and spontaneous thromboembolism has not been reported. Despite this, our case illustrates that a CPSVCA enlarges when the central venous pressure is raised. It is probably true that they do not produce pressure symptoms or rupture spontaneously because of the low systemic venous pressure. A low incidence of spontaneous thromboembolism is to be expected, since the blood flow inside the aneurysm, fusiform or saccular, is not sluggish as demonstrated by the normal transit time on radioisotopic angiocardiology.^{5, 6} However, venous thromboembolism remains a possible hazard.⁵ In conclusion, we feel that the following three points deserve consideration: (1) a saccular aneurysm should be excised, since this can be performed readily and a prosthetic graft is not required; (2) at operation, facilities for pulmonary embolectomy should be available and the aneurysm should not be manipulated until it has been excluded from the systemic venous circulation; (3) if a patient with CPSVCA is being managed conservatively, anticoagulation should be considered to prevent thromboembolism.

References

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