Schwannoma in anterior mediastinum with massive pericardial effusion

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SUMMARY

Schwannoma is a benign nerve sheath neoplasm of schwann cell origin. Mediastinum is the second most common site of schwannoma with majority originating in the posterior mediastinum. Mediastinal schwannoma is slow growing and mostly asymptomatic in presentation. We present a case of schwannoma arising in the anterior mediastinum, an unusual location, complicated by massive haemorrhagic pericardial effusion. We recommend an aggressive approach in such cases with pericardiectomy to prevent reaccumulation of the pericardial fluid.

BACKGROUND

Benign schwannomas are the most frequently encountered type of neurogenic tumour of the mediastinum located mostly in the posterior compartment.¹⁻² Mediastinal schwannomas are slow growing, typically benign and usually pose no immediate risk. We report a case of schwannoma arising at an unusual site the anterior mediastinum that was complicated by massive haemorrhagic pericardial effusion.

CASE PRESENTATION

A 15-year-old presented with history of low-grade intermittent fever for 1 month. The patient also complained of progressive breathlessness, swelling of both lower limbs and dull aching central chest pain for 15 days.

General examination revealed pulse 94/min, regular; blood pressure 90/60 mm Hg, respiratory rate 24/min; temperature 100.2°F; mild icterus; engorged neck veins and bilateral pedal oedema. Findings of cardiovascular system examination showed impalpable apex beat with muffled heart sounds and no obvious murmur. Respiratory system examination showed left-sided diminished breath sounds with dullness in the left infrascapular region and fine crackles in the right infrascapular and the bilateral interscapular area. Abdominal examination revealed soft tender hepatomegaly (3 cm below subcostal margin) and free fluid.

INVESTIGATIONS

Haemoglobin 13.4 g/dl, total leucocyte counts 12 000/mm³ (polymorphs 54% and lymphocytes 46%), platelet count 2.82×10^5 /mm³, blood urea 42 mg/dl, serum creatinine 1.10 mg/dl, serum bilirubin 1.6 mg/dl, alanine aminotransferase 86 IU/l, aspartate aminotransferase 84 IU/l and alkaline phosphatase 334 IU/l. In x-ray chest posteroanterior view, there was cardiomegaly with small left-sided pleural effusion (figure 1). In electrocardiography, there were low-voltage complexes. Two-dimensional

ECG showed large pericardial effusion. Pericardial fluid examination revealed haemorrhagic fluid with total leucocyte counts 990 cells/mm³ (polymorphs 10%, lymphocytes 70% and mesothelial cells 20%), protein 4.4 g/dl, albumin 2.7 g/dl, sugar 70.6 mg/dl, adenosine deaminase 50.5 IU/l. Contrast-enhanced CT thorax showed bulky and heterogeneous anterior mediastinal mass with bilateral pleural effusion and pericardial effusion (figure 2).

DIFFERENTIAL DIAGNOSIS

- ▶ Thymoma
- ▶ Mediastinal germ cell tumour-teratoma, seminoma
- ▶ Mediastinal goitre
- ▶ Mediastinal parathyroid adenoma
- ▶ Hodgkin's lymphoma and non-Hodgkins lymphoma

TREATMENT

Initially, pericardiocentesis was carried out and about 1.8 litres of haemorrhagic fluid was drained. Considering the biochemical parameters of the pericardial fluid, antitubercular therapy with steroids (prednisolone 20 mg twice daily) was started. The patient showed some improvement symptomatically, but on removal of the catheter, the fluid again rapidly reaccumulated in the pericardial sac. Repeat pericardiocentesis was carried out but again the fluid reaccumulated. The patient was then referred to the cardiothoracic vascular surgery department for surgical intervention where excisional biopsy of the anterior mediastinal tumour was carried out under general anaesthesia and pericardiectomy was also conducted to prevent reaccumulation of pericardial fluid.

The biopsy report revealed benign spindle cell neoplasm arranged in fascicles, intermixed with

Figure 1 Chest x-ray posteroanterior view showing cardiomegaly.

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Figure 2 CT thorax showing anterior mediastinal mass and pericardial effusion.

hypocellular and hypercellular areas (figure 3). The hypercellular areas showed a palisading pattern with formation of verocay bodies while the hypocellular areas showed dispersed spindle cells in the myxoid stroma. Immunohistochemistry examination showed that the sample was S 100 antigen positive and desmin negative suggestive of schwannoma (figure 4).

OUTCOME AND FOLLOW-UP

Early pericardiectomy with excision of tumour mass led to successful outcome.

DISCUSSION

Nearly 45% of schwannomas occur in head and neck with 9% occurring in the mediastinum.³ Mediastinal schwannomas most frequently arise in a paravertebral location from sympathetic trunks or intercostals nerves.⁴ The initial presentation with fever, breathlessness and evidence of large pericardial effusion was suggestive of tuberculosis which is the commonest cause in our country in such cases. That is why initially he was given antitubercular treatment with steroids but did not respond. Besides tuberculosis, other important causes of haemorrhagic pericardial effusion to be considered are trauma, malignancy, uraemic pericarditis, following myocardial infarction, rheumatic fever and anticoagulant therapy. There was no evidence for any of these conditions in the patient.

Mediastinal schwannomas are slow growing and mostly asymptomatic and are rarely complicated by massive pericardial effusion. There are few reported cases of schwannoma with cardiac involvement. Two of the cases are reported with pericardial effusion^{5 6} and three other with cardiac tamponade.⁷⁻⁹

Although mediastinal schwannomas are usually benign, this case suggests that they can sometime cause life-threatening complications and thus should be managed aggressively and completely resected, once discovered to prevent life-threatening complications. Thus, this case presented with an uncommon disorder in an unusual location with an unusual presentation.

Learning points

- ▶ Schwannomas are the most common neurogenic tumour of the mediastinum located mostly in the posterior compartment.
- ▶ They are slow growing, typically benign and usually pose no immediate risk.
- ▶ Mediasinal schwannoma can rarely present in anterior compartment with life-threatening complications.
- ▶ Timely diagnosis and appropriate management may prevent such complications.

Figure 3 Histological section showing loosely scattered spindle-shaped schwann cells intermixed with foamy macrophages (H&E×200).

Figure 4 Immunohistochemistry showing S100 positivity in tumour cells (Immunostain S100×200).

Competing interests None.

Patient consent Obtained.

REFERENCES

- 1 Strollo DC, Rosado-de-Christenson ML, Jett JR. Primary mediastinal tumors: part II. Tumors of the middle and posterior mediastinum *Chest* 1997;112:1344–57.
- 2 Fierro N, Del Morelli A, *et al.* Posterior mediastinum neoplasms: a case of schwannoma. *G Chir* 2004;25:35–8.
- 3 Das Gupta TK, Brasfield RD. Benign solitary schwannomas (neurilemmomas). *Cancer* 1969;24:355–66.
- 4 Marchevsky AM. Mediastinal tumors of peripheral nervous system origin. *Semin Diagn Pathol* 1999;16:65–78.
- 5 Rausche T, El-Mokthari NE, Kruger D, *et al.* Benign mediastinal schwannoma; cardiac considerations—case report and a short review of the literature. *Clin Res Cardiol* 2006;95:422–4.
- 6 Preusse CJ, Schulte HD, Irlich G. Hemodynamic cardiovascular effects caused by mediastinal tumors. *Langenbecks Arch Chir* 1986;369:149–52.
- 7 Bennis A, Ridai M, Nafidi S, *et al.* Malignant mediastinal schwannoma in Recklinghausen's disease presenting as cardiac tamponade. Reports of two cases. *Arch Mal Coeur Vaiss* 2000;93:381–4.
- 8 Soulami S, Chraïbi S, Ait Ben Hammou C, *et al.* Mediastinal malignant schwannoma disclosed by pericardial tamponade in von Recklinghausen disease. *Ann Cardiol Angeiol (Paris)* 1995;44:418–21.
- 9 Kato M, Shiota S, Shiga K, *et al.* Benign giant mediastinal schwannoma presenting as cardiac tamponade in a woman: a case report. *J Med Case Reports* 2011;5:61.

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