CASE REPORT

Mediastinal cavernous haemangioma in a patient with Klippel-Trenaunay syndrome

P-H Kuo, Y-C Chang, J-H Liou, J-M Lee

The Klippel-Trenaunay syndrome (KTS) is a rare syndrome characterised by the triad of varicose veins, bony and soft tissue hypertrophy, and cutaneous haemangiomas. A 30 year old man with KTS with a right mediastinal mass which progressively enlarged over 5 years is described. Computed tomography, magnetic resonance imaging, and bronchial angiography revealed a vascular lesion in the azygous area. After complete excision of the mass, histological examination revealed cavernous haemangioma. To our knowledge, this is the first report of intrathoracic haemangioma in KTS.

Klippel-Trenaunay syndrome (KTS) is a rare congenital generalised mesodermal abnormality characterised by the triad of varicose veins, cutaneous haemangiomas, and hypertrophy of soft tissue and bone. It can be diagnosed on the basis of any two of the above three features. Since the first description of this syndrome in 1990 approximately 140 cases have been published in the literature, but its aetiology remains unknown.

Previous reports of thoracic involvement in KTS were limited to pulmonary embolism, venous varicosity, and lymphatic obstruction. We present a 32 year old man with KTS with an unusual presentation of a mediastinal cavernous haemangioma in the azygous area. To our knowledge, this is the first report of intrathoracic haemangioma in KTS.

Figure 1  Chest radiograph showing a bulging lesion over the right mediastinal border (arrowheads).

CASE REPORT

A 30 year old man presented at our outpatient clinic in July 2001 complaining of intermittent cough with scanty sputum for 1 year. There were no other respiratory symptoms such as chest pain, haemoptysis, or dyspnoea. He was a heavy smoker who had consumed one pack of cigarettes a day for 10 years. He was noted to have a haemangioma over his right back and hypertrophy of the right upper extremity 17 years before this admission. The diagnosis of KTS was made at that time and the haemangioma was surgically removed. A mass in the right scrotum was noted 13 years earlier and the surgical biopsy also revealed a haemangioma. A chest radiograph at a local hospital 5 years previously was normal.

On examination the blood pressure was 130/90 mm Hg, his pulse rate was 90/min, and his respiration 25 breaths/min. There was hemihypertrophy of his right upper extremity. Chest auscultation did not reveal abnormal breathing sounds or bruits. There was no cyanosis of digital clubbing. A chest radiograph at this time showed a mediastinal lesion over the right perihilar area (fig 1); a computed tomography (CT) scan of the chest showed a mass with only partial enhancement located between the superior vena cava (SVC) and azygous vein (fig 2); and magnetic resonance (MR) imaging (fig 3) revealed a lesion low in signal intensity (SI) on the T₁-weighted image and high in SI on the T₂-weighted image, with intense enhancement after gadolinium diethylene-triamine penta-acetic acid (Gd-DTPA) injection. Angiography showed a hypervascular tumour in the right paratracheal area which was supplied by the right bronchial artery.

The patient underwent a thoracotomy with en bloc excision of the vascular tumour. The tumour was found firmly adhered to the SVC, trachea, and azygous vein. The blood supply of the tumour came from the bronchial vessels, and the bridging arch between the azygous vein and SVC was obliterated by the

Figure 2  CT scan of the chest revealing a mass with only partial enhancement located between the superior vena cava and the azygous vein.
hypertension or right ventricular failure in patients with KTS.

The vascular disorder in KTS is a combined capillary, venous, arteriovenous malformations seen in the Osler-Weber-Rendu syndrome, epithelioid haemangioendothelioma, sclerosing haemangioma, haemangioepicytomas, angiolipoma, lymphangiomata, and thymolipoma (especially the fibrous variant).

A cautious approach is warranted in children and young adults with mediastinal haemangiomas because some lesions have been known to regress spontaneously. CT scanning and MR imaging are helpful in identifying the extent of visceral and soft tissue involvement. For lesions that are symptomatic or that compromise vital structures, surgical excision remains the treatment of choice.

We have described an adult with KTS with a progressively enlarged cavernous haemangioma. Physicians should be aware of this rare disease entity. Close observation of patients with KTS for internal vascular anomalies is necessary to prevent life threatening complications such as pulmonary thromboembolism or compression of vital structures.

REFERENCES

Mediastinal cavernous haemangioma in a patient with Klippel-Trenaunay syndrome

P-H Kuo, Y-C Chang, J-H Liou and J-M Lee

Thorax 2003 58: 183-184
doi: 10.1136/thorax.58.2.183

Updated information and services can be found at:
http://thorax.bmj.com/content/58/2/183

These include:

References
This article cites 9 articles, 0 of which you can access for free at:
http://thorax.bmj.com/content/58/2/183#BIBL

Email alerting service
Receive free email alerts when new articles cite this article. Sign up in the box at the top right corner of the online article.

Topic Collections
Articles on similar topics can be found in the following collections
Radiology (diagnostics) (812)

Notes

To request permissions go to:
http://group.bmj.com/group/rights-licensing/permissions

To order reprints go to:
http://journals.bmj.com/cgi/reprintform

To subscribe to BMJ go to:
http://group.bmj.com/subscribe/