A giant thymolipoma

Charles Philip Carapinha, MB BCh, MRCS
Department of Paediatric Surgery, University of the Witwatersrand, Johannesburg

Linda Wainwright, MB BCh, FCPaed
Department of Haematology and Oncology, Chris Hani Baragwanath Hospital, Soweto

Jerome Alexander Loveland, MB BCh, FCS (SA), Cert Paed Surg
Department of Paediatric Surgery, Chris Hani Baragwanath Hospital

Corresponding author: C Carapinha (ccarapinha@hotmail.com)

A giant thymolipoma

Thymolipomas are rare benign tumours, constituting one of the differential diagnoses of an anterior mediastinal mass. These tumours may run an indolent, asymptomatic course, often achieving massive dimensions before presentation. Once symptomatic, respiratory symptoms predominate. Diagnosis should be confirmed radiologically during the pre-operative work-up, with pathognomonic features demonstrated on both computed tomography (CT) scan and magnetic resonance imaging (MRI). Open surgical resection remains the treatment of choice for lesions of a significant size, and sternotomy is well tolerated in the paediatric population.

A 10-year-old girl presented to our paediatric medical service with a short history of non-productive cough and shortness of breath. She had no significant past medical or surgical history, nor were any constitutional symptoms noted. No ptosis, exophthalmos, generalised lymphadenopathy, or features of superior vena caval obstruction were noted on general examination. Respiratory examination revealed decreased air entry bilaterally. The remainder of the physical examination was normal. At this point, a chest radiograph demonstrated an anterior mediastinal mass (Fig. 1).

Haematological and serological investigations, including tumour markers (alpha-fetoprotein, lactate dehydrogenase, beta human chorionic gonadotrophin, and alkaline phosphatase) (alpha-FP, LDH, beta HCG, ALP) were normal and not contributory to the diagnosis. Fine-needle aspiration identified primitive T cells. However, immunophenotypic analysis of a peripheral blood sample revealed no evidence of primitive T-cell populations. A bone marrow aspirate and trephine was normal. A computed tomography (CT) scan (Fig. 2) demonstrated a massive anterior mediastinal fatty tissue mass with islands and strands of soft tissue within it. No calcification was noted. The radiological appearance was pathognomonic of a thymolipoma. Mass effect on the pulmonary parenchyma was noted; however, there was no evidence of either tracheal or bronchial compression. The patient was subsequently transferred to our paediatric surgical service for definitive management.

A median sternotomy was performed, which revealed a massive anterior mediastinal soft-tissue mass (Fig. 3) with scattered fatty components and a well-defined border measuring 195×130×100 mm and weighing 1 170 g. While no vascular or pleural invasion was noted, both left and right phrenic nerves were adherent and required careful dissection to separate them from the tumour. The excision was uneventful and, after extubation on the table, the patient was transferred to our paediatric intensive care unit for 24 hours and thereafter to the general paediatric ward. The further postoperative course was uneventful, and the child made a rapid recovery with complete resolution of her symptoms. Histology confirmed the diagnosis of a thymolipoma.

Discussion

Thymolipoma is a rare benign tumour, found in the anterior mediastinum. Overall, thymic neoplasms comprise only 4% of all mediastinal tumours in children; only 2 - 9% of these are reported to be thymolipomas, with less than 45 paediatric cases reported to date. The first case was documented by Lange in 1916 and described as a lipoma of the thymus; the term ‘thymolipoma’ is attributed to Hall, as described in 1948. Thymolipomas can achieve massive dimensions – the largest paediatric and adult tumours weighed 2 235 g and 6 000 g respectively. Pre-operative diagnosis is the norm with well-described CT and MRI features, and typical findings include the presence of a well-demarcated anterior mediastinal fatty mass containing islands and strands of soft tissue, which are pathognomonic features. Histological findings include mature

Fig. 1. Chest radiograph showing a mediastinal mass.
case report

adipose tissue admixed with thymic tissue and Hassall’s corpuscles.

While the pathogenesis of this tumour has not been clearly elucidated, four possible theories have been postulated. Briefly, the first suggests that thymolipomas are lipomas arising from within the thymus.6 The third possibility (the ‘mixed tumour’ theory) suggests that both thymic and fatty tissue are neoplastic and contribute equally to the development of the thymolipoma.4 The fourth and final theory suggests that these tumours are a variant of thymomas, which undergo fatty degeneration.7

Of interest is the fact that myasthenia gravis, erythrocyte hypoplasia and hypogammaglobulinaemia have all been described in association with thymolipomas in the paediatric population. Whether these are true associations or coincidental findings is difficult to assess, as they are all isolated clinical descriptions.

Sternotomy remains the most common surgical approach5 and, in our experience, this incision has been found to be well tolerated in the paediatric age group, affording excellent access to the mediastinum and both hemithoraces. Patients can be extubated on the operating table or very shortly thereafter, and postoperative pain is easily controlled, allowing early mobilisation. While a single report describes a thoracoscopic resection in the paediatric age group,10 this approach may well be feasible in selected cases; the size of the thymolipoma will undoubtedly be the main limiting factor in the success of minimally invasive techniques.

Conclusion

Thymolipomas can attain massive dimensions before presentation, necessitating curative major surgery, which is well tolerated in the paediatric population.

References