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Case Report Open Access

A Rare and Interesting Case of Mediastinal Fibromatosis

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Abstract

Mediastinal fibromatosis is a rare entity affecting individuals of all age groups, and is extremely rare in children. Fibromatosis can occur anywhere in the body; most commonly in the superficial soft tissues, extremities, or abdomen. Bilateral pleural and mediastinal fibromatosis extending to neck along the tissue planes has not previously been reported in the literature. Most fibromatoses are benign but 10-20% can undergo malignant transformation. These tumours are initially asymptomatic, but can present with compressive symptoms as they grow in size. We present a rare case of fibromatosis of neck and mediastinum which developed in a 4 year old boy who had undergone cardiac surgery 2 years previously. Diagnostic dilemma and challenges in surgical removal are outlined.

This report documents an extremely rare case of anterior mediastinal fibromatosis, desmoid type that occurred immediately after cardiac surgery in a young child.

Keywords: Fibromatosis; Mediastinum

Case Report

A 4-year-old boy presented with a two-year history of a non-tender slow growing mass on the anterior aspect of his neck in the region of the thyroid. Neck examination revealed 5 × 5 cm fixed, painless, stony hard mass on the anterior aspect of neck extending to the sterno-clavicular joint; lower extent of which could not be palpated, suggestive of retrosternal extension. Swelling was neither moving with deglutition nor with protrusion of tongue. He had undergone surgery for mitral valve regurgitation 2 years previously, following which he developed this swelling. Physical examination showed signs of respiratory distress, i.e., noisy breathing with use of accessory muscles of respiration. His voice was normal but according to his parents he would develop severe stridor at night which was audible even at 20 metres. There was a vertical scar 11 cm long, along the sternum from his previous cardiac surgery. Fibre optic Laryngoscopy revealed palsy of right vocal cord with restricted movement of left vocal cord.

Investigations

Blood tests serum alpha-fetoprotein, serum beta-human chorionic gonadotropin hormone levels, and 24-hour vanillylmandelic acid levels, which were within normal limits. Serum LDH levels were raised i.e., 556.

Chest radiographs revealed a large mass extending from neck to chest.

Computed tomography (CT) showed homogenously enhancing mass extending from the hyoid bone to the upper border of the heart with bony erosions along the sternoclavicular joint. There was severe compression of the trachea with the narrowest lumen being 2.5 cm above the carina and with an antero-posterior diameter of 2 mm. significant tracheamalacia was also noted on comparisons of inspiratory and expiratory phases. No evidence of intratracheal extension or intratumoral calcification was detected.

Magnetic resonance (MR) imaging (1.5 T, Signa system) showed that the mass was slightly hyper intense relative to adjacent muscle on both T1-weighted and fast spin-echo T2-weighted images. Mass was homogenous and line of demarcation was seen with heart but not with lung pleura (Figures 1 and 2).

Fibreoptic laryngoscopy revealed palsy of right vocal cord with restricted movement of left vocal cord.

Fine needle aspiration cytology showed spindle cells with lymphocytes suggestive of Thymoma or germ cell tumour.

Biopsy of the mass was done, and microscopic examination revealed fragments of tumour tissue composed of bundles of fibroblasts suggestive of fibromatosis.

Differentials: Thymic tumors and Germ cell tumours.

Treatment

Patient underwent surgery for excision of mass through trans-cervical and midline sternotomy approach. Intraoperatively, 10 × 11 cm well-defined mediastinal mass was seen with investment of the medial ends of clavicle and superomedial part of sternum and extending to the lung pleura, pericardium, aorta, right brachiocephalic vein, bilateral common carotid arteries, Left brachicephalic vein, phrenic nerve and trachea. En bloc excision of the mass consisted of midline sternotomy, preservation of left recurrent laryngeal nerve. Mass was densely adherent to left brachiocephalic vein and since there was no plane for dissection, the left brachiocephalic vein was ligated and chest drains were inserted bilaterally (Figures 3 and 4). The surgical specimen appears to be well-demarcated, pinkish-white mass relatively avascular mass with gritty hard consistency. Microscopic examination revealed interlacing fascicles of spindle-shaped fibroblasts and an abundance of collagen without any mitotic activity. The final diagnosis of mediastinal fibromatosis, desmoids type was confirmed (Figure 5). The patient remained intubated for 3 days. He developed Left arm and facial oedema in postoperative period immediately which subsided completely within 3 days and chest drains were removed on 5th post-operative day. He had noisy breathing but was not desaturating. Clinically breathing

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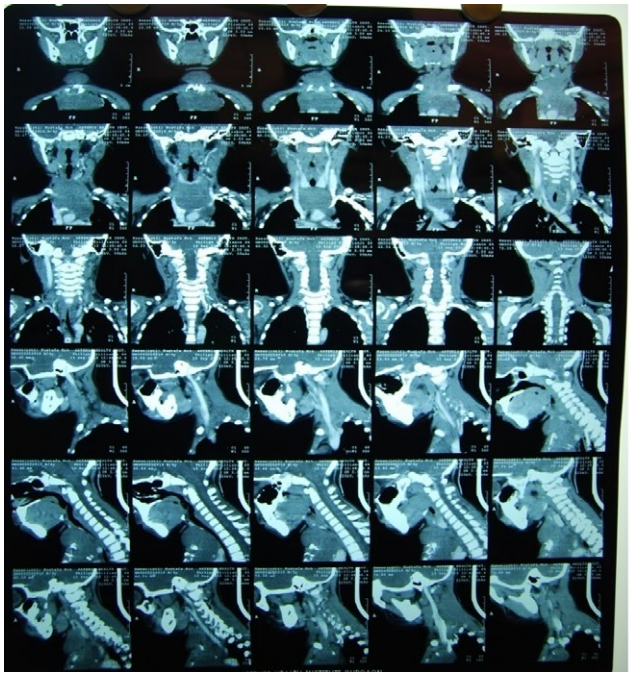

Figure 1: Scan of patient with extent of tumour.

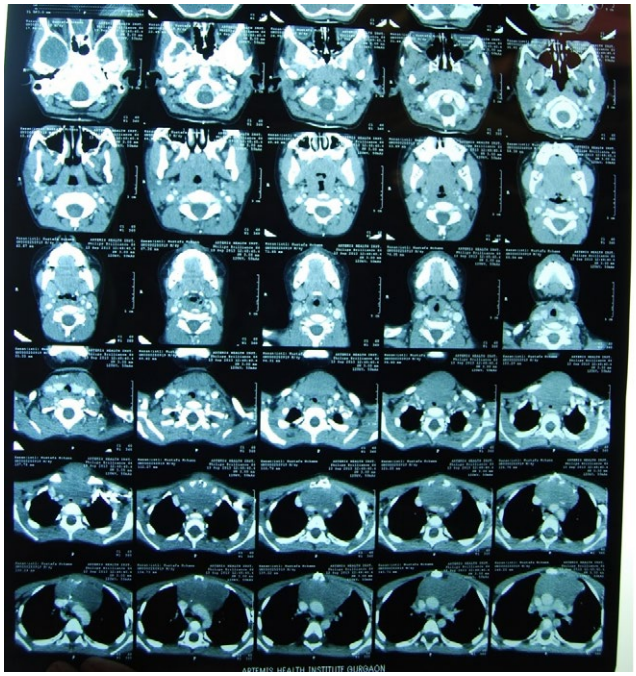

Figure 2: Scan of patient with extent of tumour.

pattern was not associated with chest retraction and tachycardia. However stridor increased progressively till 14 day post operatively. Fibre optic laryngoscopy revealed right vocal cord palsy and flicker of movement along left cord. Patient underwent right vocal cord lateralization with posterior cordotomy under general anaesthesia after 1 month.

Outcome and follow up - Presently the child does not have stridor and has acceptable voice. He has cough on feeding high volume liquids due to minimal aspiration. Neither adjuvant radiation therapy nor chemotherapy was instituted, and the patient will be followed up clinically as follow up CT at 6 weeks showed no signs of mass.

Discussion

The fibromatoses are a diverse group of non-metastasizing fibro proliferative tumours that are locally invasive and often recur after excision. Histologically, these tumours are characterized by fascicles of proliferating fibroblasts in a dense collagenous stroma with little or no mitotic activity [1-6].

Mediastinal fibromatosis is also termed as aggressive fibromatosis and desmoid tumour. It is a rare entity that affects patients of all age groups. It consists of dense and encapsulated collagenous tissue and highly differentiated fibroblasts. Despite its benign nature, it infiltrates surrounding tissues and may surround or compress mediastinal structures such as the aorta, trachea, oesophagus and heart [7]. In our case the tumour was found to infiltrate trachea, CCA, Left brachiocephalic artery, phrenic nerve and bilateral lung parenchyma.

Of all neoplasms, 0.03% is abdominal fibromatosis, which occur

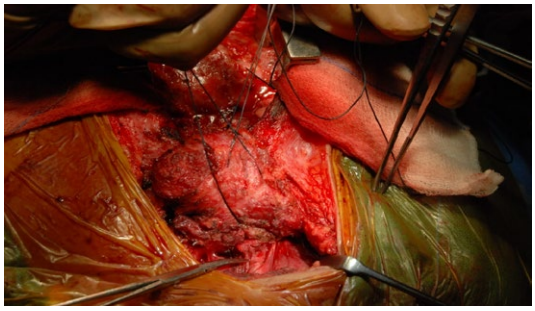

Figure3: Intraoperative findings.

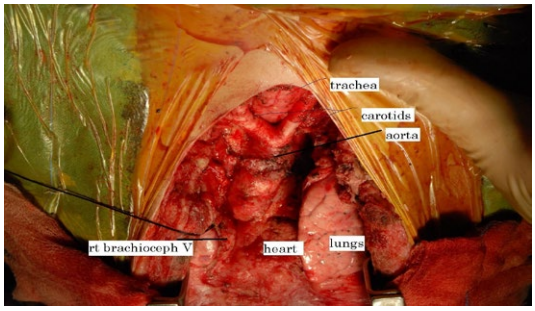

Figure 4: Intraoperative findings.

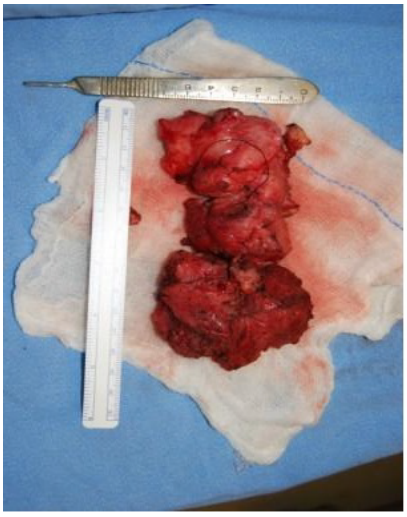

Figure 5: Specimen in toto.

most often in young women. The incidence of extra-abdominal fibromatosis was cited in 1991 as three to four cases per 1 million people [4], and we know of one similar previous case (from 900 000 children) in which anterior mediastinal fibromatosis occurred after a median sternotomy for repair of a ventricular septal defect [1].

Despite the various subgroups of adult or juvenile and superficial or deep fibromatosis that have been described, fibromatosis in children is most commonly subdivided into congenital and juvenile forms [2-4,6,8,9]. The congenital form is usually widespread with multiple destructive or infiltrative lesions that involve various bones and visceral organs and often lead to early death. The less aggressive juvenile form has a better prognosis and is usually confined to the musculoskeletal system [8,9].

Imaging features are nonspecific and biopsy is required for diagnosis. Fibromatosis usually appears on CT as a large, slow growing and infiltrative soft tissue mass. It is most frequent in the posterior and middle mediastinum but may occur in any location. On T1-weighted MRI, the mass may appear iso-intense or slightly hyper intense relative to the muscles. On fast SE T2-weighted images it appears hyper intense relative to the muscle. Enhancement occurs after intravenous administration of gadolinium [7].

Wide surgical resection with adequate margin is the treatment of choice for fibromatosis. Surgery may be difficult and is directed at relieving the compression or obstruction of vital mediastinal structures. Fibromatosis is notorious for its high rate of local recurrence but metastasis is rare. Chemotherapy and radiation therapy have been reported to reduce the rates of local recurrence [10]. However, neither treatment was appropriate for our young patient because we wanted to avoid the risks of infertility and radiation-induced secondary malignancy. Therefore, we decided to proceed with continued clinical follow-up. Differential diagnosis of this condition includes fibrosing (sclerosing) mediastinitis and well differentiated fibrosarcoma [7].

The prognosis of fibromatosis is generally quite favourable, with a 5-year survival rate of more than 90% [11], but the outcome seems to be dependent on the location of the tumour. Prompt diagnosis at an early stage is crucial, and adequate excision should be attempted in such cases.

Conclusion

Fibromatosis should always be considered as a possibility while dealing with an intrathoracic/mediastinal tumour.

The disease is potentially curable if diagnosed early.

Long term follow-up is strongly recommended for early detection of recurrence.

Conflict of Interest

Authors have no conflict of interests to declare.

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