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Sclerosing mediastinitis in a child-A case report

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Sclerosing mediastinitis is a rare non-malignant condition characterized by fibrous tissue proliferation in the mediastinum. It could be primary or secondary. It typically presents in adulthood. Sclerosing mediastinitis is extremely rare in the pediatric age group. The extensive fibrosis of mediastinal soft tissues may cause compression of the mediastinal vasculature, airway or oesophagus. Possible treatment avenues are medical, surgical resection and local therapy for complication. We report a six year old child, weighing 15 kilograms, who presented to the emergency department with respiratory collapse. Endotracheal intubation was done with a cuffed 3 mm tube because the trachea was found to be slit like and would not accommodate a larger tube. Contrast CT (Computerized Tomography) scan showed a homogenous non-enhancing mass in the anterior mediastinum and neck, obliterating the normal mediastinal fat planes and compressing the distal 3 cm of the trachea. There was no vascular compromise or esophageal compression. CT guided biopsy was reported as? thymoma. The child was taken up for excision biopsy/debulking. The intra operative finding was a very hard mass in the anterior mediastinum extending to the neck. Subtotal resection was done. The child was extubated on the first post-operative day and discharged on the fifth day. Histo-pathological examination was reported as sclerosing mediastinitis. This was a histological surprise. The child was then evaluated for tuberculosis, fungal infection, retroperitoneal fibrosis and autoimmune disease. All were negative. She returned a month after surgery with a massive right pleural effusion. Pleurocentesis was done twice. Fluid analysis did not help in diagnosing the etiology. The fluid rapidly reaccumulated. Tube thoracostomy was done and Methylprednisolone begun. The quantum of fluid decreased, but continued to drain. Antituberculous therapy was empirically begun. The patient did well thereafter. Etiologically, most cases of Sclerosing mediastintis are idiopathic/primary or secondary. Secondary cases are caused by: (1) Infection (tuberculosis, fungal infection-commonest being histoplasmosis), (2) concurrent intra-thoracic malignancy-commonest being Hodgkin's disease, (3) sarcoidosis, (4) radiation therapy and (5) drugs like methysergide association could be Riedel's thyroiditis, retroperitoneal fibrosis, autoimmune disorders such as systemic lupus erythematosus and rheumatoid arthritis, sclerosing cholangitis. The pathological types are: focal-80% and diffuse-20%. It is slowly progressive and hence asymptomatic for a long time. The clinical presentation could be systemic symptoms (fever, weight loss) or due to airway involvement (stridor, dyspnoea, wheezing, post obstructive pneumonia and atelectasis), vascular involvement (Superior vena cava syndrome, pulmonary hypertension, arterial or venous infarct) or esophageal involvement (dysphagia, esophago-bronchial fistula). Imaging modalities used are CT scan or MRI (magnetic resonance imaging). It has an unpredictable course. The three possible avenues for treatment are: Medical-anti tubercular or anti-fungal and corticosteroids, surgical resection and local therapy for complications. Prognosis depends on etiology and the structures involved.

Biography

Rekha Matta has independently performed more than ten thousand cardiovascular and thoracic surgeries (adult & pediatric). She is currently working as a professor and HOD of Department of Cardiovascular and thoracic surgery at Krishna Institute of Medical sciences deemed university at karad, India.

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