IgG4 Associated Mediastinal Fibrosis

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Introduction:

Internal jugular vein thrombosis was first described in 1912. Mediastinal compression syndrome due to internal jugular vein thrombosis (IJVT)is rare but potentially fatal condition. It usually arises following trauma to the internal jugular vein but also seen association with coagulopathies and advanced malignancies as part of para –neoplastic syndrome. Internal jugular and superior vena cava may get thrombosed due to compression in diffuse mediastinal fibrosis. The aim of anticoagulation therapy for IJVT is to inhibit further thrombus formation and prevent embolization. Thrombolysis either systemic or intracatheter is needed to in severe cases to maintain the patency of great veins and improve symptoms and complications. We present a case of IJVT due to mediastinal fibrosis where intracatheter thrombolysis resulted in almost complete patency of great veins.

Case presentation:

We report this interesting case of a 33-year old man who presented with mediastinal compression syndrome. His condition started in 2009 when he presented with three month history of progressive exertional dyspnea and was found to have constrictive pericarditis on investigations. He underwent total pericardiectomy. Pericardial histology showed evidence of granuloma but AFB stain, PCR and tissue culture for mycobacterial tuberculosis were negative. He was given empirical anti-TB treatment for 9 months. He remained asymptomatic until four year later when started to have dry cough and dyspnea on exertion. A chest X-ray showed speculated right hailer mass and right upper lobe infiltrates. CT scan of chest revealed mediastinal fullness with right hilar mass and right upper lobe infiltrates. In addition there was right para-vertebral opacity and pleural thickening. Bronchoscopy did not show any endobronchial lesion and bronchoalveolar lavage was negative for any infectious etiology. Transbronchial biopsy showed only fibrotic material with no evidence of malignancy or infection. Surgical biopsy of hilar mass and CT guided core biopsy of paravertebral mass showed inflammatory infiltrates with background of fibrosis. A final diagnosis of fibrosing mediastinitis was established after multidisciplinary meeting. Intensive workup and special staining of biopsy indicated IgG4 related disease as the cause of fibrosing mediastinitis. Patient was started on Steroids and on follow up after three months showed significant improvement both clinically and radiologically. His steroids were tapered off after a year. He remained asymptomatic until a year after discontinuing steroids when he presented with progressive shortness of breath and neck swelling. Clinical examination revealed possibility of superior vena cava obstruction in

view of engorged neck vein and facial swelling. A contrasted CT upper body and head showed extensive thrombosis of internal jugular veins bilaterally which extend superiorly to sigmoidal sinus and inferiorly to reaching the superior vena cava. In addition there was worsening of mediastinal fibrosis compressing the superior vena cava. The patient was started IV heparin infusion. In view of worsening mediastinits he was given steroids and initiated on rituximab. After a detailed venogram evaluation of great veins, intracatheter thrombolysis was done and a stent placed in superior vena cava at the site of occlusion due to mediastinal fibrosis. Subsequent venogram confirmed significant resolution of internal jugular thrombi. His symptoms improved and he was discharged on warfarin.



