

Giant calcifying fibrous pseudotumor of the neck – a case report

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ABSTRACT

Calcifying fibrous Pseudotumor is a recently recognized, uncommon benign fibrous lesion characterized by dense hyalinized collagenous tissue interspersed with benign spindle cells, psammomatous or dystrophic calcification and a lymphoplasmacytic infiltrate. It is a rare lesion with only 3 cases reported in neck region and the largest size reported in neck is of 6 cm. We report here in a case of giant calcifying fibrous Pseudotumor in the anterior aspect of neck as a first case in world literature of size more than 11cm. Although rare, calcifying fibrous pseudotumor should be included in differential diagnosis when calcific foci is identified on imaging studies in a neck mass. FNAC in such lesions are inconclusive. Radiological investigation though play a supportive role in reaching the diagnosis but final diagnosis can only be reached through histopathological examination along with immunohistochemistry.

KEY WORDS: Pseudotumor, Psammomatous, Calcification, fibrous lesion.

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Introduction

Pseudotumor is the term used for broad category of tumor proliferations that are believed to be the result of a reactive process rather than of a neoplastic nature and which can simulate malignant tumor [1, 2]. It is basically applied to Inflammatory Pseudotumor (IMT) [2]. Calcifying fibrous pseudotumor is also believed to be a pseudotumour [1]. Calcifying fibrous pseudotumor is a recently recognized rare benign lesion characterized by dense hyalinized collagenous tissue interspersed with benign spindle cells, psammomatous or dystrophic calcification and a lymphoplasmacytic infiltrate. It is

an unusual lesion that commonly occurs in soft tissue (extremities, trunk, scrotum, neck, and axilla) in young patients. Newly described locations include the chest wall, pleura, peritoneum and mediastinum [3, 4].

In our review of world literature, we found 3 cases of calcifying fibrous Pseudotumor reported in neck region. All these three cases reported in neck were of smaller size, the largest reported case was of size less than 6cm [5]. We report here a rare case of calcifying fibrous Pseudotumor in the anterolateral aspect of neck as a first case in world literature of size more than 11cm.

Case report

A 23 year old man presented to the department of otorhinolaryngology at Safdarjung hospital in New Delhi with a slowly growing swelling over the right side of the neck for the past 5 years. There was no history of pain, change of voice, dysphagia, stridor and weight loss. There was no history of trauma or surgery. His personal and medical history was unremarkable.

Clinical examination detected a firm, well defined, non-tender swelling of size approximately 12 x 9 cm² presented on the right anterolateral aspect of neck extending from lower border of mandible to suprasternal notch; and from midline to posterior

border of sternocleidomastoid [Figure-1]. The swelling was mobile anteroposterior and in vertical direction. It was not moving with tongue protrusion or deglutination and was neither translucent nor fluctuant. It did not increase in size on coughing. No bruit was heard on auscultation. Rest of ENT examination was unremarkable. The results of all routine laboratory tests were within normal limits along with thyroid function test. Fine needle aspiration cytology of the swelling was inconclusive. Ultrasound neck showed large isohypoechoic lesions in right anterior triangle of neck with multiple punctuate calcification.



Figure 1: Pre-operative photograph of the patient (right lateral view).

jugular vein was compressed by the lesion. MRI neck also confirmed the lesion to be of similar size as that of CT scan and also revealed that it is a well-defined mass on right side of neck below mandible till upper border of clavicle. Mass was displacing submandibular gland anterosuperiorly, reaching floor of mouth and medially displacing entire larynx to off midline. The mass was heterogeneously hypo intense on T-2 weighted image [Figure: 2 A&B].

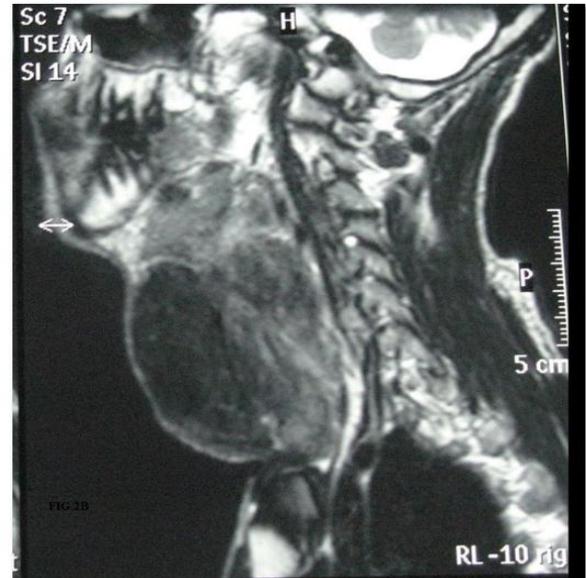


Figure 2 B: T2W sagittal images show mass to be composed of predominantly septate hypointense areas more prominent than T1W consistent with its predominant fibrotic nature.

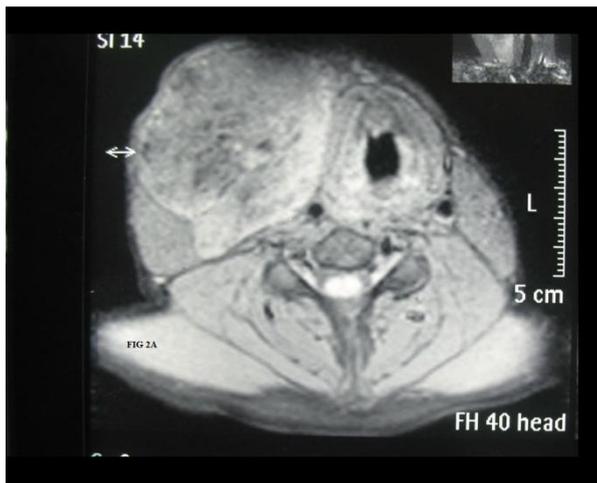


Figure 2 A: T1W fat saturated axial image shows a well-defined iso to hyperintense mass in right side lower neck region abutting right side of thyroid cartilage and right sternocleidomastoid muscle. Few hypointense areas are noted within it consistent with fibrotic/calcified foci.

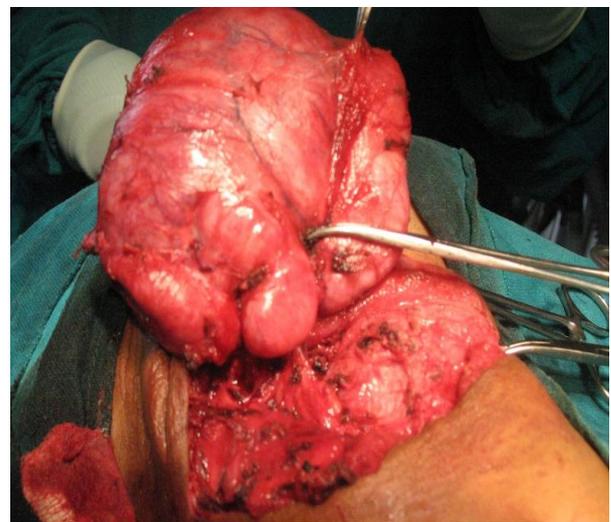


Figure 3 A: Intraoperative photograph showing tumor

Contrast enhanced computed tomography of the neck demonstrated an oval swelling of size 11 x 7.5 x 7.5 cm on right side of neck from angle of mandible to thoracic inlet with few calcific foci. The swelling was deep to platysma and right internal



Figure 3 B: Excised specimen in toto.

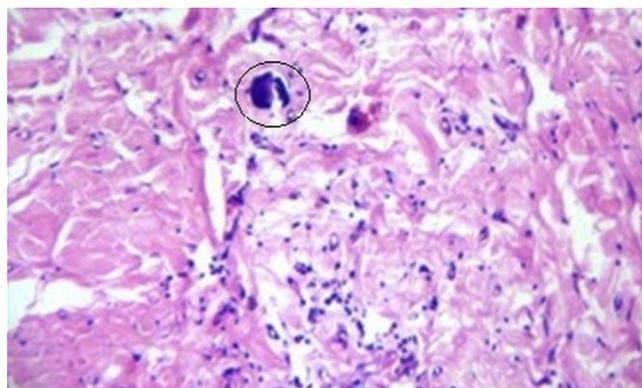


Figure 4 B: H&E 10X showing calcifications (encircled)

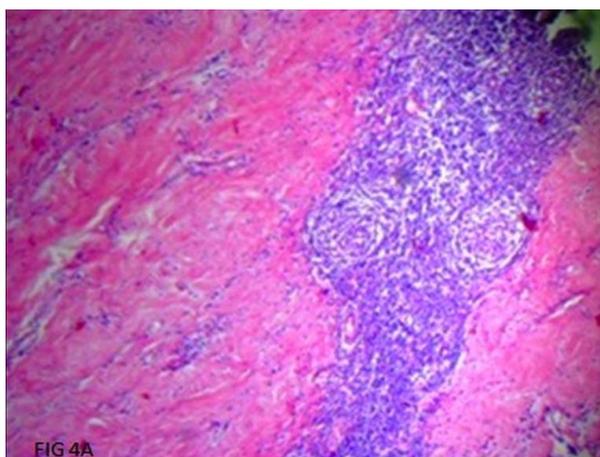


Figure 4 A: H&E 10X showing dense lymphoid aggregates in hyalinised collagenous tissue.

An excision of the mass was done under general anesthesia. A neck crease incision in the middle of the neck from anterior border of right sternocleidomastoid to anterior border of left sternocleidomastoid given. Subplatysmal flap elevated superiorly up to mandibular margin and inferiorly up to clavicle. Mass was dissected all around and was found adherent to the carotid sheath, which was freed through careful dissection. On gross examination, it was a well circumscribed, non-encapsulated, lobulated, firm nodule measuring 11 X 7 X 7 cm [Figure: 3A&B], which was removed in toto. Cut section showed homogenous grey white surface with focal haemorrhagic and calcified areas. Histological examination showed a circumscribed, paucicellular neoplasm characterized by dense eosinophilic hyalinized collagenous tissue with scattered lymphoplasmacytic infiltrate with lymphoid follicles. Focal areas of calcification seen and was consistent with calcifying fibrous pseudotumor. [Figure-4 A&B]. Immunohistochemical study shows positivity for vimentin and factorXIIIa, while staining for actin,cytokeratin,desmin and CD34 was negative.

Discussion

Calcifying fibrous Pseudotumor is an uncommon lesion that occurs mainly in young population and mostly in females [3]. The lesion was first described in literature by Rosenthal and Abdul Karim as childhood fibrous Pseudotumor with psammoma bodies [6]. Fetsch et al in 1993 used the term Calcifying fibrous Pseudotumor for the first time, when they described 10 cases of this lesion in soft tissues and stated that this proliferation is basically the result of a reactive process rather than of a neoplastic nature [1]. The pathogenesis of the lesion is unknown.

The gross pathological features include a well demarcated, non-encapsulated and lobulated mass. On sectioning, it is white to pale in color with gritty consistency, as it was in our case. Microscopic features include collection of dense hyalinized collagenous tissue interspersed with benign appearing spindle cells, with an inflammatory infiltrate mainly lymphocytes and plasma cells. Eosinophils, neutrophils and mast cells are also present in variable number. Calcification are present of either dystrophic or psammomatous types. No evidence of necrosis, atypia or mitosis is seen on microscopy. In our case, the mass was grossly non-encapsulated, well demarcated and on microscopy, it showed a circumscribed, paucicellular neoplasm characterized by dense eosinophilic hyalinized collagenous tissue with scattered lymphoplasmacytic infiltrate with lymphoid follicles.

Contrast enhanced computed tomography shows central area of increased attenuation probably due to calcification as they are of higher attenuation than enhancing vessels [4], as it was evident in our case. Together histopathological report along with radiological finding serve as an important tool for the

diagnosis of calcifying fibrous Pseudotumor. Previously, Calcifying fibrous Pseudotumor were regarded as a sclerosing end stage inflammatory myofibroblastic tumor but recently distinct features between inflammatory Pseudotumor and calcifying fibrous Pseudotumor have been suggested [2]. Inflammatory myofibroblastic tumors rarely contains calcification and have a myofibroblastic proliferation varying from hyalinized acellular collagen to florid fibroblastic proliferation varying from simulating sarcoma[7,8]. Immunohistochemical study reveals Calcifying fibrous Pseudotumor is diffusely positive for factor XIIIa and negative for smooth muscle actin, muscle specific actin and CD34. On the contrary, inflammatory myofibroblastic tumor is diffusely positive for actin, variable positivity for CD34 and focal positivity for factor XIIIa [8].

Simple local excision is believed to be curative with no recurrence after surgery. However one case of local recurrence after 7.5 years of initial surgery, however author speculated incomplete primary excision as a cause of recurrence [1]. No malignant changes have been observed even in long standing cases. Calcifying fibrous pseudotumor is a rare, benign fibrous lesion of unknown pathogenesis. These have been reported in the subcutaneous tissue of trunk, axilla, extremities, mesentery and mediastinum but there are only 3 reported cases in the head and neck region.

Calcifying fibrous pseudotumor are uncommon but it should be included in the differential diagnosis of neck masses. Calcifying fibrous pseudotumor can easily be confused with other neck masses as preoperative diagnosis of Calcifying fibrous pseudotumor is difficult. FNAC in such lesions are inconclusive. Radiological investigation though play a supportive role in reaching the diagnosis but final diagnosis can only be reached though histopathological examination along with immunohistochemistry.

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