



RESEARCH ARTICLE

NECK DESMOID TUMOR (AGGRESSIVE FIBROMATOSIS): A RARE CASE

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ABSTRACT

Background: Fibromatosis or desmoid tumors are rare (constitutes less than 3% of soft tissue tumor) benign tumors of musculo-aponeurotic origin with extensive tendency of local invasion causing morbidity.

Case Presentation: We report a rare case of 30 year old male presented with left sided neck mass situated in left supra clavicular region. Ultrasonography and Magnetic resonance imaging (MRI) of neck revealed well defined soft tissue swelling of sized 7cm x 8cm x 8cm, present in left supra clavicular region. patient underwent surgical excision of mass. Histological and immunohistochemistry analysis confirmed the diagnosis of fibromatosis (desmoid tumor) of neck.

Conclusion: Fibromatosis has unique aggressive behavior. Due to lack of set protocols for its management, treatment should be individualized and multi-modality treatment including radiotherapy and chemotherapy should be considered. We expect further awareness and study of this aggressive tumor by presenting our case.

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INTRODUCTION

Fibromatosis or desmoid tumors are rare (constitutes less than 3% of soft tissue tumor) benign tumors of musculo-aponeurotic origin with extensive tendency of local invasion causing morbidity. Most common site is abdominal wall followed by intra-abdominal. Incidence of extra-abdominal desmoid tumors is less, among which involvement of head and neck region is only 7% to 15%. Therefore, we are presenting a rare case of desmoid tumor of left supraclavicular region.

Case

A 30 year old male presented to us with left sided neck mass situated in left supra clavicular region since 7 months. There was gradual progression in size of this mass over period of 7 months to attain size of approx. 8cm×8cm immobile, non-tender, firm to hard in consistency, margins well-defined while surface was smooth with limitation of movements of neck towards left due to the swelling. Extension of swelling was superiorly up to apex of posterior triangle, inferiorly to clavicle, laterally up to trapezius and medially up to the anterior border of sternocleidomastoid muscle. There were no symptoms of trachea and esophageal compression. Patient had pulmonary Koch's 1year back and had taken medication for 7 months. Familial history was not significant. Local ultrasonography of the swelling was suggestive of

9cm×6cm×11cm well defined, heterogeneous predominant hypoechoic lesion present in left supra-clavicular region with no area of necrosis and calcification. Other neck structures were unremarkable. HRCT of neck and chest revealed a 7 cm×8 cm×8 cm heterogeneous enhancing soft tissue lesions with central non-enhancing area which was present posterior to left sternocleidomastoid muscle. There was evidence of subcentrimetric sized lymphadenopathy at level II and III. MRI neck showed a left sided supraclavicular swelling of size 8cm×7cm×7cm which appeared iso-intense on T1 images and iso to hyper-intense on T2/STIR images with fewenlarged jugularlymph nodes most probably suggestive of inflammatory etiology. Left sided surgical excision of mass was performed under general anesthesia. A single hard mass of approximate size of 8cm×8cm×9cm which found to be densely adhered to deep cervical fascia and invading to scalenus anterior, medius muscle and prevertebral fascia. Vital structures in the neck were unremarkable. On histology gross specimen was brownish, and on cut surface it was whitish with fibrotic band seen throughout the mass. Microscopy showed spindle cells embedded in fibrous matrix. Tumor cells appeared bland with spindle nuclei. There was no evidence of cellular atypia and mitosis attributed to Benign Spindle Cell Lesion. To confirm the diagnosis immuno-staining was done and found to be negative for S-100 and Positive for SMA (smooth muscle actin) and MiB-1, which was consistent with Fibromatosis (Desmoid tumor). Patient had uneventful post-operative course and was discharged on 4th post-operative day. Regular follow up period up to 5 months were unremarkable and uneventful.

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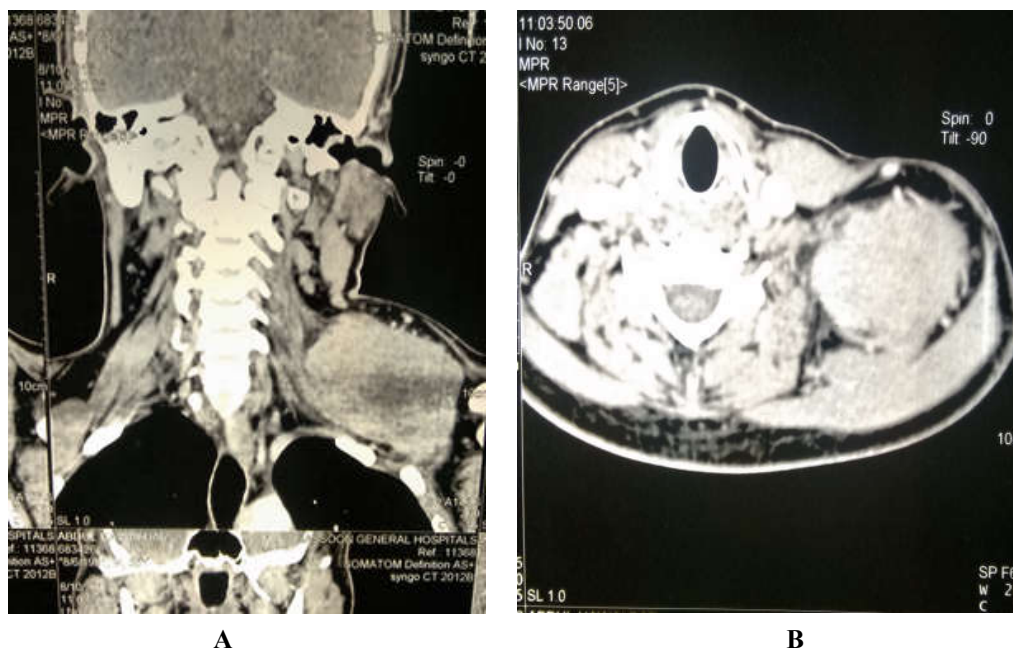


Fig. 1. HRCT neck showing mass in left suprascapular region (A) coronal (B) axial view

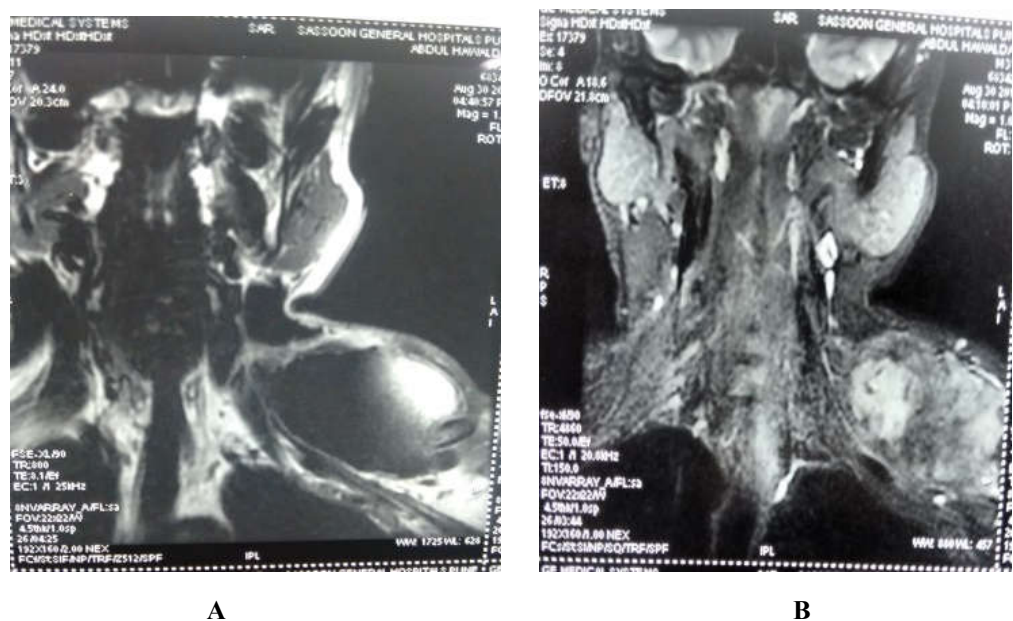


Fig. 2. MRI neck showing mass in left sided neck region (A) iso-intense on T1 and (B) iso to hyper-intense on T2 image

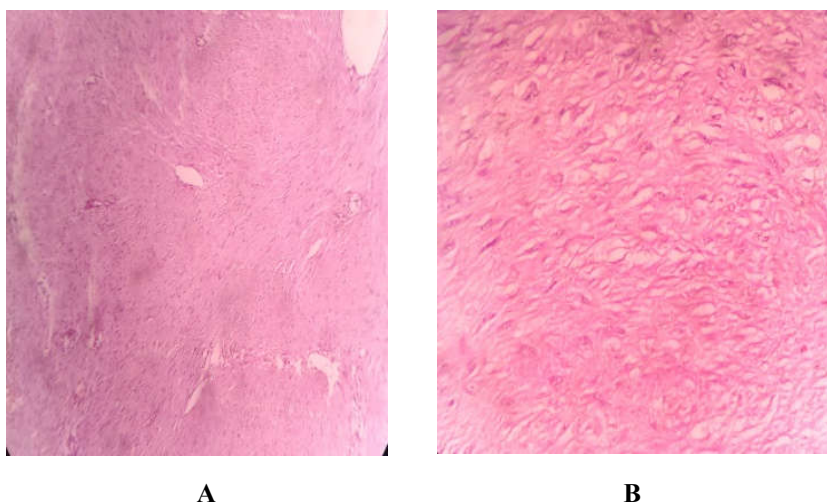


Fig. 3. H&Emicroscopy showing spindle cells embedded in fibrous stroma (A) 10x (B) 40x

DISCUSSION

Aggressive fibromatosis (Desmoid tumor) are rare, fibroblastic benign soft tissue tumor arising from musculo-aponeurotic structures throughout the body with slight female predominance with female to male ratio of 1.4:1. Desmoids are sporadic in nature with incidence of 2 to 5 cases per million per year but sometimes they do occur in patients with familial adenomatous polyposis. Most characteristic feature is local infiltration, and they do not metastasize to distant site (Lev, 2007 and Clark, 1996). There is notable lack of systematic analysis of this tumor in the literature. Aggressive fibromatosis is classified as adult type (age>20years) and juvenile type (age<20years). Adult type is further divided into two types Dupuytren and Desmoid type according to location. Desmoid tumor can be either of abdominal wall (most common), intra-abdominal or extra- abdominal. Limbs followed by head and neck are the common site for extra-abdominal desmoid tumor (Rampone, 2007). On histology, macroscopically tumor is firm, and on cut section it is greyish white while on microscopy there is interwoven bundles of spindle cells with varying amount of collagen and immunohistochemical staining positive for smooth muscle actin(SMA), as seen in this case (4). In head and neck desmoid tumor their progression is gradual as enlarging painless mass, fixed to underlying and surrounding structures. By virtue of this it may be associated with limitation of movements, trismus and pressure symptoms due to compression of trachea and esophagus (J.G. B, W. R., 1994). Although recurrence rate is higher (70%), complete excision is the only effective and standard of care for desmoid tumor.

Unlike other benign tumor of head and neck region, fibromatosis (desmoid tumor) have potential for local invasion to surrounding structures and this factor is very important in dealing with tumor in this region due to presence of complex anatomy and vital neuro-vascular structures. Due to complexity of the anatomy and nature of the tumor, sometimes complete excision of tumor is not possible and therefore aim is for an adequate local control with subsequent acceptable functional and esthetic outcome (Ballo, 1999 and R. P, A. N, R. S, 2013). Some studies favors surgery followed by adjuvant radiation therapy for residual disease control; however other studies are not in favor of effectiveness of adjuvant RT for tumor with positive microscopic margin. Radiation to head and neck region is associated with high risk of complications, therefore; this should be reserved for the non-operable conditions only (Lev, 2007; Abdelkader, 2001 and Niv, 2000). Various studies also showed use of cytotoxic and non-cytotoxic drugs. Low dose methotrexate reported to be useful in shrinkage of tumor. COX-2 inhibitor- Meloxicam has also been shown to be effective in controlling extra-abdominal fibromatosis.

Some studies also advocate the use of high dose Tamoxifen (selective estrogen receptor modulator) in combination with Sulindac (NSAIDs) after the surgical resection of the tumor for prevention of recurrence (Hansmann, 2004 and Emori, 2014). Considering this, these chemotherapy can be considered for unresectable, radiation resistant desmoid tumor and for this various trials are going on to validate their results.

Conclusion

Although Desmoid tumor is a benign neoplasm, it has unique aggressive behavior. Due to lack of set protocols for its management, treatment should be individualized and multi-modality treatment including radiotherapy and chemotherapy should be considered. Following surgical management patient should have a close observation due to unpredictable natural history of this benign tumor.

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