An Interesting Case of Fibromatosis of Neck with Review of Literature

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Received: 4 March 2013 / Accepted: 23 August 2013 / Published online: 5 September 2013
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Introduction

Fibromatosis is also known as desmoid tumour. It is a slow-growing and locally infiltrative disease caused by uncontrolled proliferation of fibrous tissue arising from deep musculoaponeurotic structures. It differs from fibrosarcoma in having low cellularity, infrequent mitoses, absence of necrosis, and not metastasising. Despite having benign histology, being locally aggressive it can cause significant morbidity [1].

Fibromatosis is a rare entity, representing only 0.03 % of all neoplasm in humans [2]. It is classified according to its anatomical position into three subtypes namely abdominal, intra-abdominal, and extra-abdominal. Fibromatosis of the head and neck region comprises around 10 to 25 % of all cases of extra-abdominal fibromatosis [3]. Attempts have been made to separate it from that arising from other parts of the body because of the unique clinical problems it can cause in relation to its critical anatomical position [3–5].

Though surgery is the main stay of therapy, multimodality treatment including partial or complete excision followed by chemo-radiation or hormonal therapy can be effective.

Case Report

A 43 yr old female presented to us with complaints of swelling in neck along with compressive symptoms in the form of

difficulty in deglutition and occasional dyspnoea since 2 months. On examination, a 17×14×10 cm neck swelling with restricted mobility was found predominantly on left side of neck (Fig. 2). There was no clinical evidence of cervical lymphadenopathy. Left carotid was palpable anterior to the swelling whereas trachea was shifted to the right side. The mass was neither moving upwards with deglutition, nor with the protrusion of tongue. Thyroid function tests were within the normal limits. X ray neck and chest were suggestive of superior mediastinal and neck mass with tracheal shift to right side. Indirect laryngoscopy was suggestive of normal movement of both the vocal cords. CECT scan of neck and chest was done which was suggestive of 17×12×13 cm sized lobulated soft tissue mass in left half of neck and supraclavicular region extending into superior mediastinum, prevertebral space and posterior triangle. Left internal jugular vein was not seen separate from the tumour and left common carotid artery was also shifted anteriorly and was partially encased by the tumour. Erosion of lateral cortices of C5-D1 vertebral bodies with pre-vertebral and para-vertebral soft tissue, extending into neural foramina between C5-D1 was also observed (Fig. 1). Preoperative cytology was done and was not conclusive of any specific disease but was suspicious of fibromatosis. Patient was assessed for retroperitoneal fibrosis which was found to be absent on USG abdomen. On exploration, the mass was densely fixed to the strap muscles of neck as well as common carotid on left side (Fig. 2). Left internal jugular was completely encased within the tumour. Meticulous dissection with partial excision of tumour with preservation of neurovascular bundle was performed. Post operative histopathological study confirmed the diagnosis of fibromatosis. Immunohistochemistry was strongly positive for actin, vimentin and desmin (Fig. 3).

Post operative period was uneventful and patient responded well to the radiation and hormonal therapy in the form of tamoxifen 20 mg twice a day for 6 months. On follow up there

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Fig. 1 CT Scan showing tumour involving left half of the neck and superior mediastinum

have been no signs of recurrence or obstructive symptoms in the past 6 years.

Discussion

Fibromatosis of the head and neck is a rare disease which usually present with a painless mass or less commonly, pain, obstructive features or neurological symptoms [4]. As with

fibromatosis arising elsewhere in the body, surgical resection is the treatment of choice [1–6]. The primary goal is to achieve a clear resection with wide margins, but preservation of function and major vessels is of equal importance. Due to the complex anatomy and frequent entrapment of neurovascular structures in the head and neck region, these aims are often difficult to achieve. Resection often leads to injury of important surrounding tissues in the form of carotid artery, internal jugular vein or brachial plexus and is associated with high

Fig. 2 Regression of tumour size after post operative tamoxifen therapy

recurrence (46–62 %) requiring repeated excisions [5, 6]. Most recurrences occur within 2 years of resection, which is the minimum time recommended for follow-up; the recurrence risk is higher for younger group of patients [4, 7]. Several series in the literature have confirmed that the margin status following resection does not necessarily correlate with local disease recurrence. Therefore we do not recommend the sacrifice of major neurovascular structures in an attempt to obtain negative margins [8].

The precise role of adjuvant therapy and therapeutic agent has not been established probably due to the rarity of the disease in head and neck. It could also be partly because of the occasional observation of spontaneous regression and arrested growth after incomplete excision, and the finding that the resection margin correlates poorly with the likelihood of local recurrence [4].

Though the efficacy of radiotherapy is controversial, there are some reports of good local control rates using radiotherapy as either adjuvant or primary therapy [8–10]. Radiotherapy should thus be employed if primary excision is not feasible, or if post operative residual tumour is remaining, or if the tumour recurs [7, 9, 10].

Hormonal agents especially anti-estrogens have also been found to be effective in fibromatosis of neck. The presence of anti-estrogen binding site (AEBS) distinct from ERs probably plays an important role in mediating the action of anti-estrogens. Furthermore, the presence of high levels of AEBS in desmoid tumours found to be ER-negative might explain the response to tamoxifen when the overall incidence and concentration of ERs is low in these tumours [11, 12]. Our patient has also responded well to the partial excision, preserving the neurovascular bundles followed by radiation and tamoxifen therapy with no signs of recurrence or any obstructive features even after a follow up period of 6 years.

Conclusion

Fibromatosis though a rare disease, should be considered a differential diagnosis of a neck swelling. Therefore, a detailed attention should be paid to all relevant clinical information. Any unusual presentation or disease course should raise suspicion for further investigations and reassessment of the case. Aggressive fibromatosis is best managed by multidisciplinary approach. The

Fig. 3 H & E along with vimentin stained tumour cells

disease is usually partially resectable when presented below the deep cervical fascia. Post operative radiation along with tamoxifen therapy offers good results when complete excision is not possible due to involvement of neurovascular structures. Follow up has to be maintained in all the cases of fibromatosis as there are high chances of recurrence especially in the first 2 years.

Conflicts of Interest There are no conflicts of interest.

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