Aggressive Fibromatosis of Head and Neck in a Child

Atul Gupta · Satish Nair · Ajith Nilakantan ·
Amit Sood

Received: 25 October 2014 / Accepted: 8 November 2014 / Published online: 13 November 2014
© Association of Otolaryngologists of India 2014

Abstract Aggressive fibromatosis is a broad group of benign fibrous tissue proliferations of similar microscopic appearance that are intermediate in their biological behavior between benign fibrous tissues and fibrosarcomas. They are rare lesions accounting for less than 3 % of all soft tissue tumours. Usual site is extremity whereas head and neck fibromatosis is a rare occurrence. We present a case of aggressive fibromatosis of the infratemporal fossa in a child, which was treated by surgical excision with adjuvant hormonal therapy.

Keywords Fibromatosis · Maxillary swing · Chemotherapy

Case Summary

6 years old boy presented with complaints of generalized swelling right side of face since 6 months of age, which has been slowly increasing in size. He also gave complaints of trismus for the past 6 months and recurrent episodes of epistaxis from right nostril for the past 4 months. There was no history of nasal blockage, headache or facial pain and difficulties in swallowing or breathing. There was also

no history of prenatal, natal or postnatal insult or trauma. On examination the child had a diffuse swelling in the right hemiface especially in the region of cheek and angle of mandible although there was no palpable lesion. Mouth opening was restricted (2 cm). There was a smooth submucosal bulge over the right side of soft palate and tonsil with the tonsil being pushed medially (Fig. 1). Nasal endoscopy revealed a smooth submucosal bulge in the lateral wall of right side of nasopharynx. Right ear showed features of otitis media with effusion. There were no cranial nerve deficits or palpable cervical lymphadenopathy.

On radiological evaluation contrast enhanced CT scan revealed a non-enhancing soft tissue density in right infratemporal fossa and parapharyngeal space. Contrast enhanced MRI neck and brain revealed a large well-defined mass ($5.7 \times 4.9 \times 3.7$ cm) in the right infratemporal fossa, pterygopalatine fossa and parapharyngeal space with its epicentre in the region of pterygoid muscles (Fig. 2). There was also associated bony destruction of the sphenoid sinus superiorly, the posterior wall of maxillary sinus anteriorly and the mandibular condyle laterally with focal intra cranial extension into the middle cranial fossa. The presumptive diagnosis was a case of soft tissue tumour in view of the extent and bony destruction but the long history suggested a benign condition.

The child was taken up for examination under anesthesia and biopsy of the lesion was obtained. The histopathological evaluation revealed spindle cells in sweeping fascicles arranged in herringbone pattern with hypercellular areas admixed with lesser-hyalinised areas. No myxoid or lipomatous differentiation was noted. Immunohistochemistry was positive for Vimentin and mitotic index in terms of Ki 67 labelled tumour cells was less than 1 % in cellular areas. The final diagnosis was Fibromatosis (Fig. 3). The case was discussed in the institutional tumour board and a

A. Gupta · S. Nair (✉) · A. Nilakantan · A. Sood
Department of Otolaryngology and Head & Neck Surgery, Army
Hospital (Research and Referral), Delhi 110010, India
e-mail: aachku@yahoo.com; aachku@gmail.com

A. Gupta
e-mail: atulrh@gmail.com

A. Nilakantan
e-mail: ajith.nil@gmail.com

A. Sood
e-mail: amitsood_afmc@yahoo.com

Fig. 1 Intraoral submucosal smooth swelling of the *right* side of soft palate

decision of surgical excision with adjuvant therapy was finalised. In view of the extensive involvement and difficult anatomy for excision, it was decided that the best chance for adequate excision would be by a maxillary swing approach.

A right Weber-Fergusson incision with subciliary extension with midline upper lip split approach was taken. The bone cuts were made at premaxilla, midline hard palate, frontal process of maxilla and the zygomatic process of maxilla after marking for titanium miniplates. Right maxillary swing was performed to identify a 6 × 5 cm, firm, greyish white irregular mass in the infratemporal, parapharyngeal and infratemporal fossa. The lesion appeared to be infiltrating the pterygoid muscles with focal area of erosion of skull base and sphenoid with no intracranial extension. The tumour was excised and the maxilla was replaced and fixed with titanium miniplates and the

Fig. 2 **a** Axial cuts of contrast enhanced CT scan revealing lesion in the *right* infratemporal fossa. **b** Axial cuts of contrast enhanced CT scan revealing lesion in the parapharyngeal space. **c** Axial cuts of contrast enhanced MRI scan revealing lesion in the parapharyngeal space. **d** Coronal cuts of contrast enhanced MRI scan revealing lesion in the parapharyngeal space-extending lateral to cavernous sinus intracranially without dural involvement

palate with an obturator (Fig. 4). Postoperative period was uneventful. The final histopathological diagnosis confirmed the diagnosis of Fibromatosis.

The child was planned for adjuvant therapy, as complete excision with adequate margins is difficult in the pterygoid region. In view of the histology, age of child and the effects of external radiotherapy at the critical area of skull base, the institutional tumour board decided to exhibit adjuvant systemic therapy with antiestrogen Tamoxifen in the dosage of 50 mg once a day and Sulindac in the dosage of 50 mg twice a day. The patient is on Tab Tamoxifen and Sulindac for past 6 months and has a stable disease state with minimal disease in the infratemporal fossa without progression as seen on follow-up radiological evaluation.

Fig. 3 Histopathology showing classical herring bone pattern

Fig. 4 **a** Weber Fergusson incision with upper lip split and subciliary extension on *right* side. **b** Flaps raised with marking of bony cuts. **c** Maxillary swing done exposing the right infratemporal fossa. **d** Tumour exposed in *right* infratemporal fossa and parapharyngeal space. **e** Titanium miniplates used to close the maxilla. **f** Skin closure

Discussion

Infantile fibromatosis (IF) is a benign, locally infiltrative fibrous tissue proliferation composed of mature fibroblasts and collagen without cellular anaplasia [1]. They were first described by Stout and represent a distinct entity of intermediate lesions arising from the musculoaponeurotic system. They are characterized by high propensity to invade adjacent structures without distant metastasis [2].

WHO classifies IF into diffuse (mesenchymal) and desmoid types [3]. The diffuse has predominantly immature fibroblasts and low collagen whereas desmoid variety as was our case has more of mature fibroblasts and collagen. The usual site of IF is anterior abdominal wall, shoulder and upper arms where they arise from the connective tissue of muscles, overlying fascia or aponeurosis. Occurrence of IF in head and neck is rare.

The exact aetiology is still unknown with various factors like genetic, viral, hormonal (oestrogen), trauma and irradiation being proposed to cause it [4]. The presence of membrane-specific hormone receptors on the fibroblasts supports a hormonal-based theory and hormonal therapy for these cases are based on these findings.

IF appear macroscopically as ill defined firm greyish white mass with a strong infiltrative growth pattern along tissue planes with an ability to invade adjacent tissues. Histologically they consist of well-differentiated fibroblast, fibrocytes and myofibroblasts with a collagenous to myxoid stroma. Atypical mitotic figures or anaplastic cells are not seen.

Radiological evaluation plays an important role in identifying the extent for preoperative planning and follow

up after treatment. CT scan provides information about bone involvement whereas MRI provides better soft tissue delineation especially in head and neck lesions with identification of intracranial involvement [5].

Surgical excision and Radiotherapy are the two principal modalities of treatment [6]. A complete en-bloc resection is the treatment of choice if possible. In head and neck, unless the lesion is small, complete resection with wide margins is technically difficult due to the complex anatomy, presence of important structures and the chances of increased morbidity after excision. Treatment response rates for tumor-free and tumor-positive margins respectively for surgery are 72 and 41 % whereas adjunctive treatment with postoperative radiotherapy improves it to 94 and 75 % [7].

Other adjunctive treatment modalities described are use of hormonal therapy and chemotherapy [8]. There are limited studies on the effectiveness of these modalities and most of the data is available as case reports or case series. The systemic therapy commonly used is the antiestrogen Tamoxifen, which causes inhibition of ER beta expression. It is found to cause partial response to the tumour or stabilizes disease process and prevents progression [9]. In addition to Tamoxifen, high dose Sulindac has also been used as it inhibits beta catenin signaling and has been found to have additive effect to Tamoxifen [9, 10]. A combination of vincristine, actinomycin D and cyclophosphamide has also been used in few selective cases not responding to hormonal therapy.

Our case due to the extensive involvement of infratemporal fossa and skull base was managed surgically with maxillary swing approach for resection. Maxillary swing is an excellent approach to the nasopharynx, pterygopalatine fossa and infratemporal fossa. It provides excellent exposure and the postoperative morbidity is minimal. In view of the critical area involved with difficulty in achieving negative margins, the patient was planned to be exhibited with postoperative adjuvant therapy. Due to the young age and the effects of external radiotherapy, patient was managed with antiestrogen Tamoxifen and Sulindac and presently has a stable disease state.

Conclusions

The aim of the report was to highlight infantile fibromatosis of head and neck region as a rare benign lesion with high propensity for local infiltration. Surgical excision with adequate margins is the treatment of choice, which can be difficult in cases of head and neck owing to the complex anatomy. Postoperative adjunctive radiotherapy and hormonal therapy improves the local control rates.

References

1. Plaat BE, Balm AJ, Loftus BM, Gregor RT, Hilgers FJ, Keus RB (1995) Fibromatosis of the head and neck. *Clin Otolaryngol* 20:103–108
2. Abdelkader M, Riad M, Williams A (2001) Aggressive fibromatosis of the head and neck (desmoid tumours). *J Laryngol Otol* 115:772–776
3. Eizinger FM, Weiss SW (1995) Fibrous tumors of infancy and childhood. In: Eizinger FM, Weiss SW (eds) *Soft tissue tumors*, 3rd edn. Mosby-Year Book, St Louis, pp 231–268
4. Hoos A, Lewis JJ, Urist MJ, Shaha AR, Hawkins WG, Shah JP, Brennan MF (2000) Desmoid tumors of the head and neck—a clinical study of a rare entity. *Head Neck* 22:814–821
5. Ahn JM, Yoon HK, Suh YL, Kim EY, Han BK, Yoon JH et al (2000) Infantile fibromatosis in childhood: findings on MR imaging and pathologic correlation. *Clin Radiol* 55:19–24
6. Wang CP, Chang YL, Ko JY, Cheng CH, Yeh CF, Lou PJ (2006) Desmoid tumor of the head and neck. *Head Neck* 28:1008–1013
7. Nuytens JJ, Rust PF, Thomas CR, Turrissi AT III (2000) Surgery versus radiation therapy for patients with aggressive fibromatosis or desmoid tumors. A comparative review of 22 articles. *Cancer* 88:1517–1523
8. Schmitt G, Mills EE, Levin V, Smit BJ, Borcker H, Pape H (1992) Radiotherapy of aggressive fibromatosis. *Eur J Cancer* 28:832–835
9. Janinis J, Patriki M, Vini L, Aravantinos G, Whelan JS (2003) The pharmacological treatment of aggressive fibromatosis: a systematic review. *Ann Onc* 14(2):181–190
10. Hansmann A, Adolph C, Vogel T, Unger A, Moeslein G (2004) High-dose tamoxifen and sulindac as first-line treatment for desmoid tumors. *Cancer* 100:612–620