Pseudosarcoma: A diagnostic and treatment dilemma

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A 11-year-old female presented with a one-month history of a rapidly growing tumor of the upper alveolus accompanied by loosening of the associated teeth. Excision biopsy was reported as “cellular spindle cell tumor”. However, the lesion recurred and in a matter of days attained its preoperative size. On examination she had a 3 cm growth on the left alveolus abutting the premolars and molars. The lesion was bright red, exophytic, firm and bled on touch. A CT scan of the paranasal sinuses showed a lytic lesion in the floor and lateral wall of the maxilla without gross involvement of the maxillary antrum [Figure 1].

After review of the biopsy slides and discussion in the cancer board, the patient underwent an inferior partial maxillectomy. Postoperatively the patient was fitted with a palatal obturator. Recovery was uneventful. There was no evidence of recurrence after one year.

The resected portion of the hard palate included a 2.9x2.5 cm growth, with a grey white cut surface [Figure 2].

Microscopically there were ill-defined whorls and fascicles of spindle-shaped cells with tapering cytoplasm and elongated nuclei [Figure 3]. The latter were vesicular with a fine chromatin pattern and small nucleoli. Nuclear hyperchromatism and pleomorphism were absent. Cellularity was generally moderate as a result of separation of the cells by edema. However, a few foci were more solidly cellular. Scattered mitotic figures were observed but no atypical forms could be found. There was a prominent vascular network in the lesion, some parts of which had a “tissue culture” appearance that resembled granulation tissue. The presence of an edematous and richly vascular tissue culture pattern combined with the absence of nuclear atypia, abnormal mitotic figures and a well developed fascicular pattern constituted reassuring evidence that the lesion was indeed
benign and the features seen best fitted nodular fasciitis-a pseudosarcomatous lesion.

Nodular fasciitis is an entirely benign but rapidly growing lesion. This condition was first described by Konwaler in 1955.[1] It is believed to be an exaggerated reparative process of uncertain cause.[2] These lesions are commonly seen in the extremities in adults. Growths resembling nodular fasciitis have been described at visceral sites, especially in the genito-urinary tract.[3] A variety of names have been applied to these lesions including inflammatory pseudotumor, pseudosarcomatous fibro- myxoid tumor, postoperative spindle cell nodule and nodular fascitis.[1] Whether these visceral tumefactions are identical to nodular fasciitis of the somatic tissues is uncertain, but there are many similarities.[3] Nodular fasciitis involving the maxilla appears to be very rare with only one more case reported in the English literature.[4]

The proliferating cells are of fibroblastic and myofibroblastic type. The high cellularity and mitotic activity can mimic sarcoma. It is especially problematic in the pediatric population in which nodular fasciitis is not commonly encountered while mesenchymal malignancies of the head and neck are of fundamental concern. The alternative possibilities are leiomyosarcoma, fibrosarcoma and even sarcomatoid carcinoma. The rapid clinical onset, presence of stromal chronic inflammation and lack of cytological atypia are all critical features that help identify the lesion as benign.[5]

Local excision is the treatment generally advocated for nodular fasciitis but intra-lesional steroids have been tried with varied success.[6] Spontaneous regression of these lesions is also known to occur.[3] Owing to its ominous presentation nodular fasciitis has a tendency to be over-treated, as in the present case. In hindsight, curettage and intra-lesional steroids may have been sufficient and would also have avoided a major surgical procedure.

References


Source of Support: Nil. Conflict of Interest: None declared.