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OLGU SUNUMU

Fibromatosis of the mandible in a child

Çocukta mandibüler fibromatozis: Olgu sunumu

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Fibromatosis represents a group of fibrous tumors showing clinic and biologic features between benign fibrous lesions and fibrosarcoma. These locally aggressive tumors have high recurrence rates (20% to 70%). A four-year-old boy presented with mandibular fibromatosis occupying the mandible completely and extending to the submandibular gland and soft tissues. Complete hemimandibulectomy and submandibular gland excision were performed followed by reconstruction with a curved Kirschner wire. No signs of recurrence was observed during a follow-up period of 18 months. In addition, no limitations in the functions of the jaw, mastication, and swallowing were noted.

Key Words: Biopsy; child, preschool; fibroma/surgery; fibroma, desmoplastic/surgery; fibromatosis, aggressive/surgery; head and neck neoplasms/surgery; mandibular neoplasms/surgery.

Fibromatozis, benign fibröz lezyonlar ve fibrosarkom arasında klinik ve biyolojik özellikler gösteren fibröz tümörler grubudur. Lokal olarak agresif bir seyir izleyen bu tümörlerin rekürens oranları da yüksektir (%20-70). Dört yaşında bir erkek çocukta mandibulu tümüyle dolduran, submandibüler gland ve yumuşak dokulara yayılım gösteren fibromatozis saptandı. Tedavi olarak hemimandibulektomi ve submandibüler gland eksizyonu uygulandı ve Kirschner teli ile rekonstrüksiyon yapıldı. On sekiz aylık takip süresince herhangi bir rekürens gözlenmedi. Çene, çiğneme ve yutma fonksiyonlarında kısıtlılık görülmedi.

Anahtar Sözcükler: Biyopsi; çocuk, okul öncesi; fibroma/cerrahi; fibroma, desmoplastik/cerrahi; fibromatosis, agresif/cerrahi; baş-boyun neoplazmları/cerrahi; mandibüler neoplazmlar/cerrahi.

Fibromatosis represents an interesting group of fibrous tissue neoplasms falling between benign fibrous proliferations and fibrosarcoma of unknown etiology[1] and consisting of well-differentiated fibroblasts with no atypical mitosis or anaplastic elements.[2]

There are different kinds of fibromatosis; they present similar histological features, but diverse clinical behaviours, leading to various definitions and classification systems.[1,3] Desmoplastic fibroma represents the intraosseous counterpart of soft tissue fibromatosis or desmoid tumor, in that its histologic appearance is identical to that of the extraosseous desmoid. These lesions have also been reported as fibromatosis or juvenile fibromatosis of bone.[4]

They may be locally aggressive, invade surrounding structures and recur after resection. These nonen-

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capsulated lesions originate from fascia, musculoaponeuroses or the periosteum. Although they do not exhibit metastases, they have a tendency for recurrence ranging from 20% to 70%.[3] The incidence of fibromatosis is 2.4 to 4.3 per million individuals per year.[5,6] In the maxillofacial region, it is most commonly encountered in the posterior mandible. The choice of treatment is usually surgical resection of the lesions showing infiltration into soft tissues.[1,7]

In this case report a case of mandibular fibromatosis is presented.

CASE REPORT

A four-year-old boy was referred to our clinic with a history of painless enlargement of the right mandible. Limitation in the opening of the mandible was noted. The patient did not have paresthesia or anesthesia of the inferior alveolar nerve distribution. Clinical examination showed an immobile and non-tender swelling of the right mandible, extending to the submandibular gland (Fig. 1). No palpable lympadenopathy was observed in the neck. Intraoral examination showed a slightly bulging mass in the vestibule. General physical examination showed no abnormality.

X-ray radiograms revealed a well-demarcated, radiolucent lesion. Computed tomographic examination of the mandible showed an expansive radiolucent lesion measuring 5x4x4 cm, with a lytic process extending from the condyle posteriorly to the canine tooth anteriorly (Fig. 2).

An incisional biopsy was performed under general anesthesia and histologic examination revealed fibromatosis. Because of the rarity of the lesion site, frozen section examination was also performed during surgery. Nearly complete occupation of the mandible with tumor from the mentum to the condyle and its wide extension into the submandibular gland and soft tissues necessitated complete hemimandibulectomy and submandibular gland excision instead of simply local excision and curettage. An immediate and transient reconstruction with a curved Kirschner wire was performed to obtain mandibular symmetry without any restriction in growing (Fig. 3, 4).

Macroscopically, the tumor filled the medullary cavity of the mandible causing thinning and perforation of the cortex in several areas. There was infiltration to the submandibular gland. Histologically, the tumor exhibited features identical to those observed in the biopsy specimen with no cytologic atypia (Fig. 5). No signs of recurrence was observed during a follow-up period of 18 months. In addition, no limitations in the functions of the jaw, mastication, and swallowing were noted.

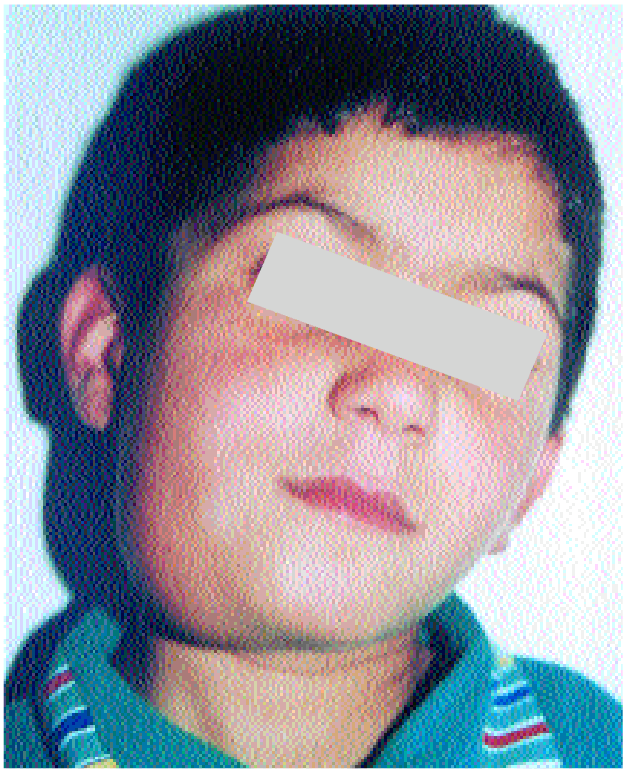

Fig. 1 - *Immobile and non-tender swelling of the right mandible, extending to the submandibular gland.*

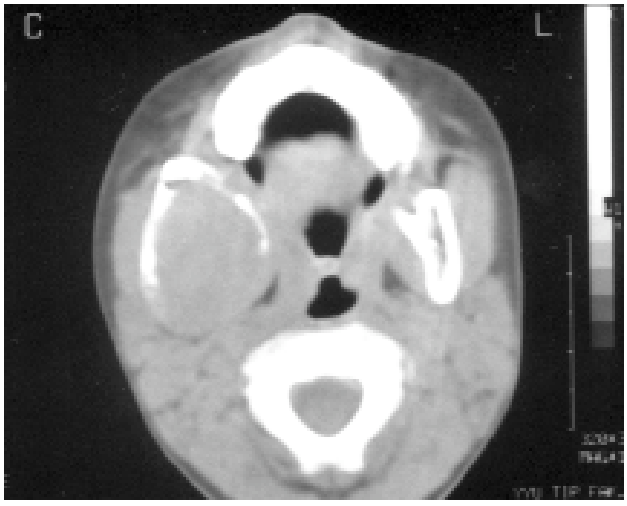

Fig. 2 - *Computed tomography view of the mandible showing an expansive radiolucent lesion with a lytic process.*

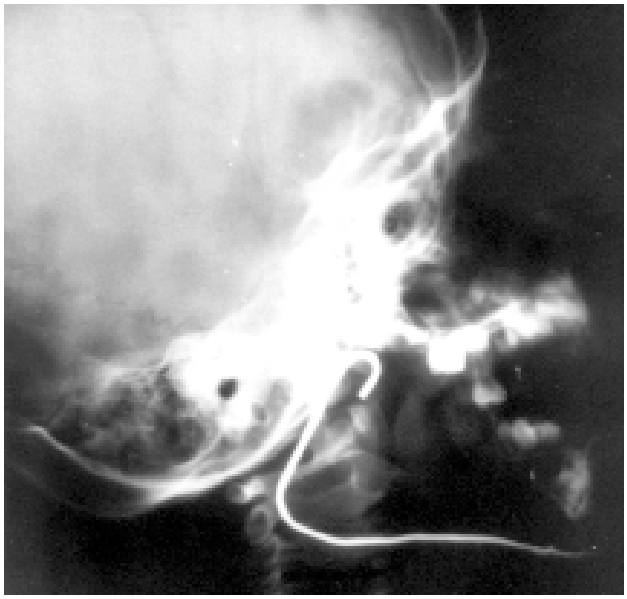

Fig. 3 - *Reconstruction of the defect was performed with the use of a curved Kirschner wire.*

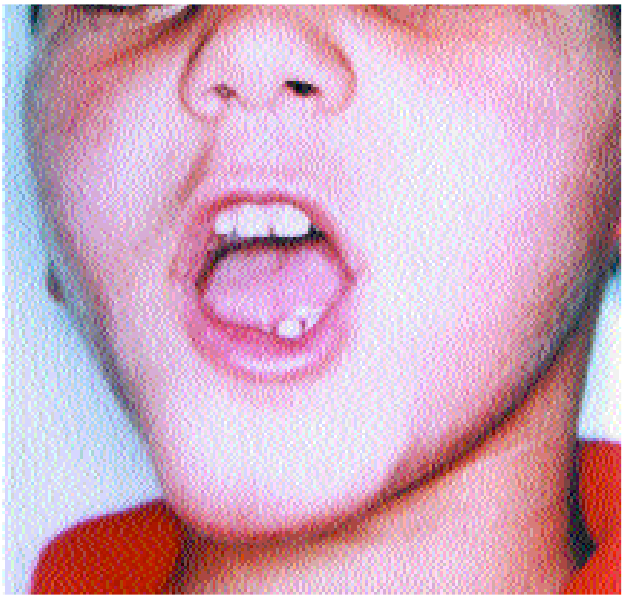

Fig. 4 - *A symmetric facial appearance was obtained after post - operative two months.*

DISCUSSION

Although fibroma usually involves the long bones, it may rarely occur in the mandible,[8] or even more rarely seen in the maxilla.[9] Differential diagnosis includes myxoma, neurofibroma, xanthofibroma, reostal desmoid, nasopharyngeal fibroma, chondromixoid fibroma, and congenital generalized fibromatosis.[8] The etiology of fibromatosis is still controversial.[3] Histologically, the center of the tumor is cellular, but the periphery often shows vigorous cellularity that can be mistaken for low grade fibrosarcoma. As seen in our case, the mandibular molar-ramus-angle region is the most common maxillofacial site involved.[3] The absence of an encapsulating envelope might explain the potential extension to the surrounding structures. In a review of 26 patients, the lesions exhibited erosion of the cortical plate in 77%, and extension into the muscle in 42%.[9] In our case, we also found erosion on the cortical plate, and extension into the submandibular gland and soft tissues, which

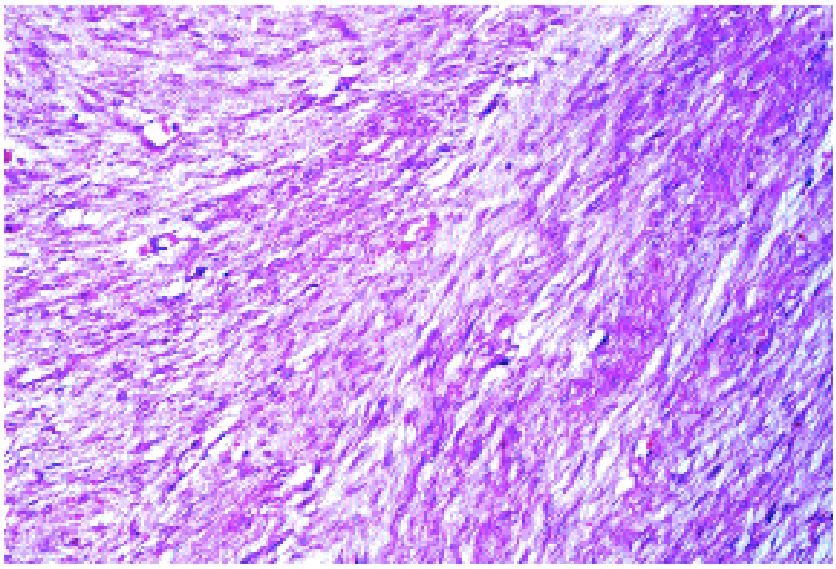

Fig. 5 - *Arrangement of tumor cells in ill-defined fascicles (H-E x 50).*

required complete hemimandibulectomy and submandibular gland excision.

As to the treatment, there have been reports of good response with chemotherapy and radiotherapy.[10,11] Raney et al.[11] reported three patients treated with VAC regimen (vincristine, actinomycin D, and cyclophosphamide), two of whom showed complete disappearance and one had 50% regression. In addition, antiestrogen treatment may be helpful in patients with estrogen receptor positive tumors.[12,13]

Controversy exists regarding the best method of surgical treatment. Segmental resection or even curettage can be carried out. Freedman et al.[9] found that, of 22 patients with desmoplastic fibroma, 19 had no evidence of recurrence throughout a follow-up period of three months to eight years. He concluded that curettage was more appropriate until further long-term studies showed otherwise. We believe that the preferred method of treatment should be relied upon the agressiveness of the lesion. En bloc resection of the tumor and excision of surrounding structures with a wide margin of bone seems to be the most effective initial treatment.

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