*Case Report*
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# A Submandibular Fibromatosis; A Case Report and Review of Literature

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## Abstract

**Background:** Fibromatosis is a group of benign tumors originating from connective tissues of muscle, overlying fascia, periosteum, or aponeurosis. However, they might need several excisional resections, owing to compressive effects on adjacent vital structures.

**Case Report:** Here, we discussed a case of submandibular fibromatosis in a 3-year-old girl. She underwent a conservative surgical procedure without a wide mandible bone resection. She received no adjuvant therapy. The postoperative period was uneventful with an appropriate cosmetic and functional outcome. A 1-year follow-up revealed no recurrence.

**Conclusion:** Fibromatosis of the head and neck in children is a rare condition, which needs a multidisciplinary agreement for its appropriate management. A complete surgical removal often leads to a proper outcome. Adjuvant therapy should also be kept in mind for recurrent lesions not candidate for surgical removal or in case of remnant tumors.

**Keywords:** Desmoid Tumor, Fibromatosis, Submandibular, Surgical Excision

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## Introduction

Desmoid fibromatosis is a rare, benign tumor that usually originates from the connective tissue with a high propensity for local recurrence (1). It has an unpredictable course and might progress or subside, which makes the management challenging (2). The World Health Organization (WHO) defined desmoid tumors as a "clonal fibroblastic proliferation that arises in the deep soft tissues and is characterized by infiltrative growth and a tendency toward local recurrence, but an inability to metastasize."(3) Its overall incidence is about 5 cases in a million of the general population annually (4). This case report aimed to discuss diagnostic and therapeutic aspects of submandibular fibromatosis in a 3-year-old girl.

## Case Presentation

A 3-year-old girl was presented to our hospital with a history of progressive enlargement of the right submandibular region over the past 4 months. Her parents reported no history of previous trauma and no obvious pain, difficulty in swallowing or breathing, fever, and weight loss. Her medical and family history was unremarkable. The detailed physical examination revealed a 4 × 3 centimeter nontender right submandibular mass fixed to adjacent structures (Fig. 1). No obvious skin changes were evident. The neurologic examination and review of systems revealed normal results. Panoramic and lateral radiographs of the mandible showed a soft tissue density with adjacent bone spike-like periosteal reaction (Fig. 2).

Results of serial axial computed tomography of the mandible were suggestive of a well-defined enhancing soft tissue mass measuring 36 × 37 mm adjacent to the body of the mandible on the right side (Fig. 3). The mass seemed to originate from the mylohyoid muscle and displaced the genioglossus muscle to the opposite side and submandibular gland posteriorly. Bony erosion and periosteal reaction alongside a regional lymph node with a short-axis diameter of 9 mm were noted. Multiple sublingual, submandibular, parotid and posterior cervical lymph nodes (maximal short-axis diameter of 12 millimeters) were also detected.

Magnetic resonance imaging of the neck revealed a soft tissue mass in the right submandibular region measuring 4.2 × 3.1 cm. A focal area of increased intensity in the bone marrow of the right mandible was seen and the right

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*Fig. 1.* Frontal view of the child showing the right submandibular mass

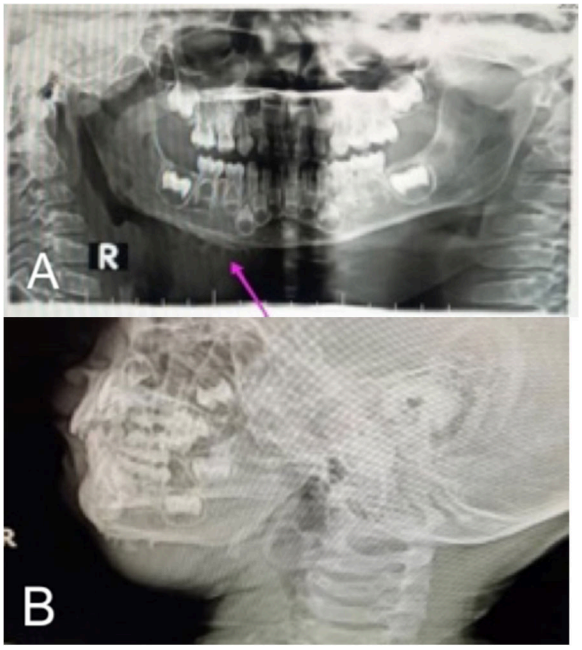


*Fig. 2.* A. A panoramic view of the mandible. B. A lateral view of the mandible revealing soft tissue density alongside with spike like periosteal reaction

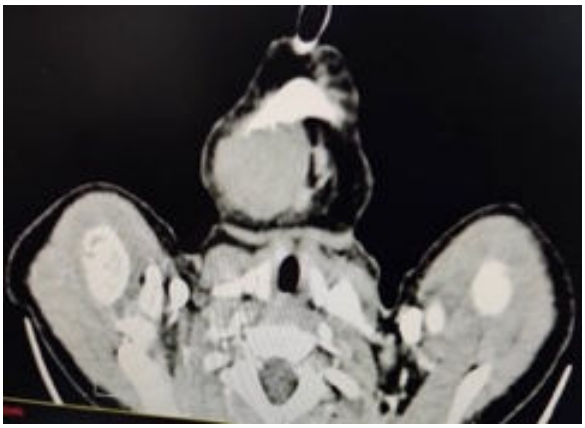

*Fig. 3.* Computed tomography showing a well-defined enhancing mass of the right submandibular area

mandible was encased by the mass. The right submandibular gland seemed separate from the mass. Multiple cervical lymph nodes were noted.

Biopsy of the mass revealed spindle cell mesenchymal neoplasm with mild atypia consistent with either fibromatosis, infantile myofibroma, or infantile fibrosarcoma. In a microscopic study, 1 to 2 mitoses per high power field were seen but no necrosis was reported. Immunohistochemical staining showed positive staining for vimentin, Beta-catenin, CD34, and SMA. Ki67 was expressed in 2% to 3% of cells. Staining for desmin, S100, P53, CD68, and CD117 had negative results. The IHC result was compatible with fibromatosis.

A surgical plan was discussed with her parents and informed written consent was obtained. Under general anesthesia, a 3-cm incision was made beneath the angle of the mandible and the flap was elevated in the subplatysmal plane. The right submandibular gland was resected. The mandible was exposed in a subperiosteal plane. The mass and the muscles of the floor of the mass that seemed involved were resected. Marginal mandibulectomy was performed and the submental and marginal mandibular nerves were preserved (Fig. 4). The wound was closed in 2 layers and a compressive dressing was applied. The postoperative 1-year follow-up was uneventful and showed no sign of recurrence or obvious mandible deformity (Fig. 5).

The final histopathologic analysis of the mass showed irregular fascicular proliferation of bland-looking spindle cells with extravascular red blood cells and little myxoid stroma (Fig. 6). No significant necrosis or mitosis was found. Immunohistochemistry staining showed positive reactivity for Beta-catenin and negative reactivity for P53, ALK, desmin, myogenin, SMA, S100, and CD34. The findings were compatible with the diagnosis of fibromatosis.

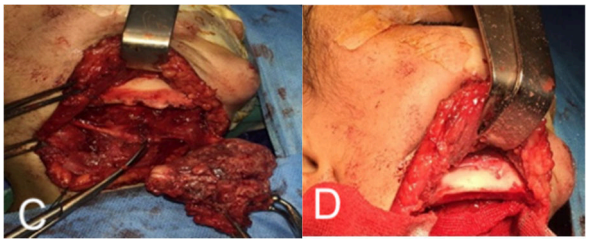


*Fig. 4.* C. Spiculated projections of the mandible alongside with the excised mass D. Mandible after marginal mandibulectomy

Fig. 5. One-year follow-up showing complete resolution



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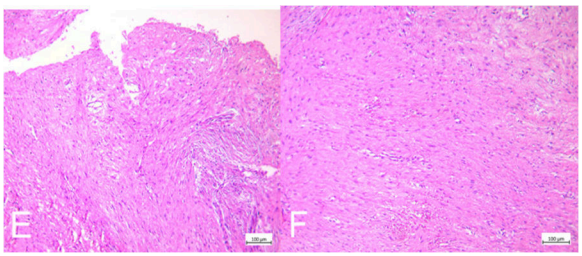


*Fig. 6.* Sections show the lesion with poorly defined borders composed of spindle cells with uniform appearance and pale cytoplasm in a collagenous stroma. No nuclear hyperchromasia but minimal cytologic atypia is seen. Hypocellular area with stromal hyalinization and thin walled vessels are also noted.

## Discussion

The fibroblastic proliferation of desmoid tumors are benign tumors with varying clinical features. Myofibroblasts are thought to be the origin of the tumors when there is no true capsule surrounding them. These tumors are found in almost all parts of the body and might present as occupying lesions. The site of the tumor determines the signs and symptoms and could be life-threatening in case of pressure on vital organs (2, 5).

Here, we reported a 3-year-old girl who presented to our hospital with a history of progressive enlargement of the right submandibular region over the past 4 months. She had no history of trauma as a precursor for the tumor growth. Despite this fact, many studies reported trauma as an underlying reason for extra-abdominal desmoid tumors in up to 80% of reported cases. For instance, Rohana Ali reported submandibular desmoid fibromatosis following a wisdom tooth extraction (6). However, there is still not enough evidence to support the role of trauma in developing fibromatosis as in our study.

The etiology of desmoid tumors is not well known, but they might be associated with some mutations in beta-catenin or APC (Adenomatous Polyposis Coli). Moreover, there is an unknown association between pregnancy and desmoid tumors, which is thought to be due to hormonal changes or a surgical incision (7). The diagnosis is based on history taking and physical examination of a soft tissue mass approved by pathological study. CTNNB1 and APC mutations are mutually exclusive in desmoid tumors and can help to exclude a syndromic condition. Therefore, it has been recommended to evaluate the mutations to confirm the diagnosis and guide further therapeutic steps (7, 8).

The treatment of choice is complete surgical resection. However, recurrence is reported in many patients, especially if a safe margin has not been achieved. A safe margin is usually recommended to be at least 2 to 4 cm (9). However, in some cases, such safe margins might not be achieved trying to preserve the function and aesthetic appearance. The head and neck area in children is among those areas in which a surgical approach is often challenging.

In our patient, because the tumor had a progressive increasing size and caused cosmetic issues, surgical resection was decided. However, her parents did not consent to a wide resection and reconstruction. Therefore, the plan changed to a free macroscopic margin rather than a microscopic one. As in some case reports, there was a need for mandibulectomy (10).

The most recent consensus guideline for the management of desmoid tumors was released in 2018 (7). The treatment approach is widely discussed in this guideline. Based on the latest guideline, a definite surgery or another form of active treatment are now considered the main treatment. If the tumor is in a noncritical site and without any symptoms, a subtle progression in the tumor size could be ignored as the course of the disease is unpredictable.

It is recommended to perform a primary magnetic resonance imaging (computed tomography, if not available) and then repeated every 3 to 6 months. Active surveillance avoids overtreatment in those patients in whom the tumor is stable. On the other hand, tumors of specific regions, such as the head and neck or mesenteric, should undergo a prompt surgical excision (7).

Additive modalities, such as radiotherapy, have also been used in the treatment of desmoid tumors in the elderly, patient intolerance, and comorbidities, alone or in combination with surgery. The risk of local recurrence is slightly lower in combination therapy but without a statistically significant difference. Furthermore, in conditions when surgery is not applicable, a moderate dose definitive radiotherapy could stop the tumor progression (11, 12).

Antihormonal therapies/NSAIDs; tyrosine kinase inhibitors (TKIs), such as imatinib, nilotinib, sorafenib, pazopani; and chemotherapy options have also been studied. However, due to the rarity of the condition, there is a lack of well-designed comparative studies and there is still no consensus. Thus, it is suggested to consider the patient's condition, overall survival, drug toxicity, availability, and level of evidence to administer these medications. Management of desmoid tumors in pediatrics is almost the same as in adults and no major differences exist (7, 13).

## Conclusion

Fibromatosis of the head and neck in children is a rare condition, which needs a multidisciplinary agreement for its appropriate management. A complete surgical removal often leads to a proper outcome. Adjuvant therapy should also be kept in mind for recurrent lesions not candidate for surgical removal or in case of remnant tumors.

### *Conflict of Interests*

The authors declare that they have no competing interests.

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