

Cervicofacial Desmoid Tumor in a Child: A Case Report

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DOI: <https://doi.org/10.36347/sjmcr.2026.v14i10.036>

| Received: 10.07.2026 | Accepted: 21.09.2026 | Published: 15.10.2026

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Abstract

Case Report

Desmoid tumor, or aggressive fibromatosis, is a benign fibrous tissue tumor characterized by a high potential for locoregional invasion; it causes significant morbidity, particularly when located in the head and neck region. We report the case of an 11-month-old infant presenting with disfiguring aggressive fibromatosis [desmoid tumor] of the right infratemporal fossa. The diagnosis was established via histological analysis of a tumor biopsy, and treatment consisted of surgical resection; however, signs of cervical recurrence appeared one year postoperatively.

Keywords: desmoid tumor, aggressive fibromatosis.

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INTRODUCTION

According to the World Health Organization [WHO] [1], desmoid tumors are fibroblastic and myofibroblastic tumors of intermediate malignancy arising from the musculoaponeurotic structures of deep soft tissues; they are characterized by a high potential for locoregional invasion and recurrence, with a predilection for the head and neck region.

Their clinical and radiological presentation is nonspecific, yet they pose a differential diagnostic challenge regarding malignant tumors due to their highly infiltrative nature.

Histological diagnosis via tumor biopsy, supplemented by immunohistochemical analysis, is essential prior to determining any therapeutic strategy.

Treatment protocols remain poorly standardized; various therapeutic approaches are employed. While surgery is the gold standard, chemotherapy, radiotherapy, and hormonal therapy are used as adjuncts or for unresectable tumors. The quality of the surgical treatment influences the likelihood of subsequent recurrence [2].

Desmoid tumors can occur at any age and can arise anywhere in the body; they are categorized into three groups based on location: intra-abdominal and

mesenteric [8%], abdominal wall [49%], and extra-abdominal [43%] [3,4].

Head and neck locations account for 12% to 15% of extra-abdominal desmoid tumors in adults [5,6]. This location is more common in children, with an estimated incidence of 22% to 35% of extra-abdominal desmoid tumors [7].

Macroscopically, desmoid tumors appear as hard, whitish, ill-defined nodules or masses that infiltrate skeletal muscle. They have a fibrous, "band-like" consistency [8].

Microscopically, they consist of a proliferation of fibroblastic cells within varying amounts of collagenous tissue [tumors ranging from more cellular to more fibrous], with infiltrative, spiculate, tentacle-like extensions. They tend to adhere to surrounding structures. Mitotic activity is usually low, and necrosis is rare [9,10].

CASE REPORT

An 11-month-old patient, born of a first-degree consanguineous union following a well-monitored, full-term pregnancy and an uncomplicated Caesarean delivery, with good adaptation to extrauterine life.

Since birth, the patient has presented with a right cheek swelling causing facial asymmetry and

progressively increasing in size; this prompted a consultation with an ENT surgeon at the age of 9 months. A biopsy of the cheek mass was performed, yielding two fragments measuring 2 mm and 3 mm along their longest axes. Histopathological examination revealed a spindle-cell proliferation composed of scattered elements and regularly interlaced bundles, showing low to moderate cellularity; the nuclei were regular with fine chromatin, and no mitotic activity was observed. Immunostaining showed strong positivity for anti-smooth muscle actin

antibody, while the cells tested negative for anti-desmin and anti-S100 antibodies.

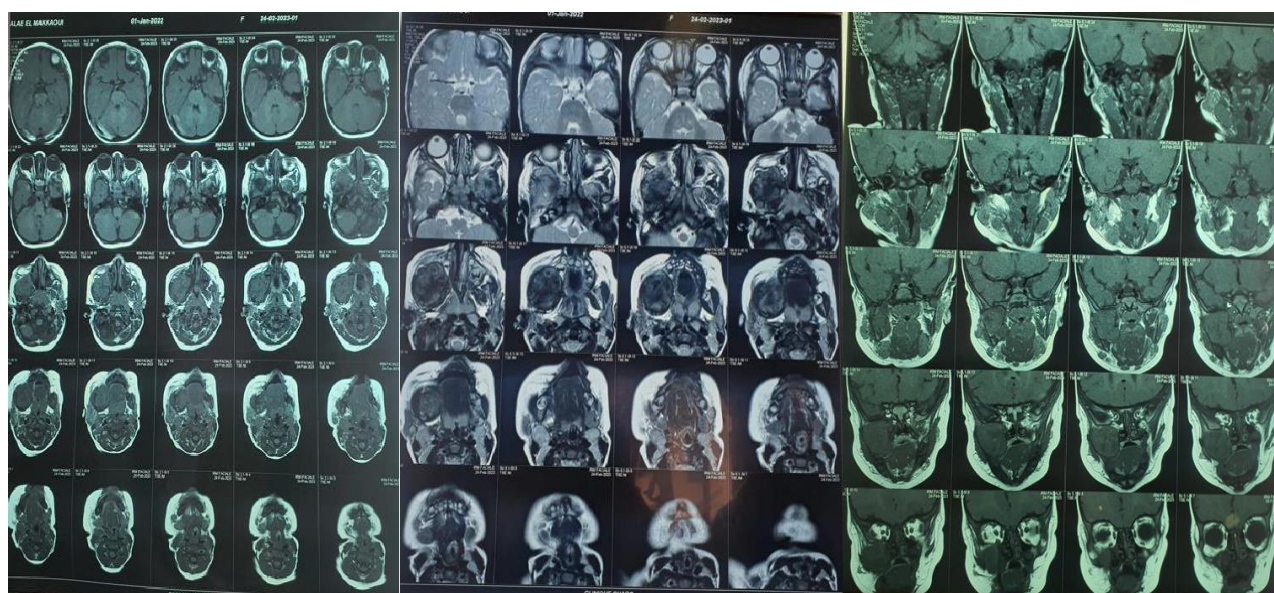
Clinical examination

Upon admission, a right cheek swelling was noted, causing facial asymmetry; it was painless, with no overlying signs of inflammation or infection and no sensory or motor deficits. Mouth opening was adequate, with a painless, budding swelling on the inner surface of the right cheek that did not bleed on contact and was associated with obliteration of the upper buccal vestibule.



Facial MRI: A tumoral process centered on the right infratemporal fossa, showing T1 hypointensity and heterogeneous, moderate T2 hyperintensity, with heterogeneous enhancement following gadolinium injection. The lesion measures 40 x 35 x 35 mm and appears well-defined; anteriorly, it fills the ipsilateral

maxillary sinus, while laterally, it invades the adjacent ascending ramus of the mandible, though the signal of the adjacent cheek soft tissues remains preserved. Anteriorly, there is intimate contact with the greater wing of the sphenoid and the floor of the right orbit, with signs of infiltration in certain areas.



Facial CT scan:

Large, relatively well-defined tumor mass centered on the right infratemporal fossa;

Irregular contours in places; tissue density showing enhancement following contrast injection; dimensions: 40 x 34 x 37 mm.

Superiorly: Causes widening of the inferior orbital fissure; abuts the inferior rectus muscle [with a visible separation plane] and the right orbital floor.

Medially: Displaces the hypoplastic right maxillary sinus; abuts the mandible, causing lysis of its outer

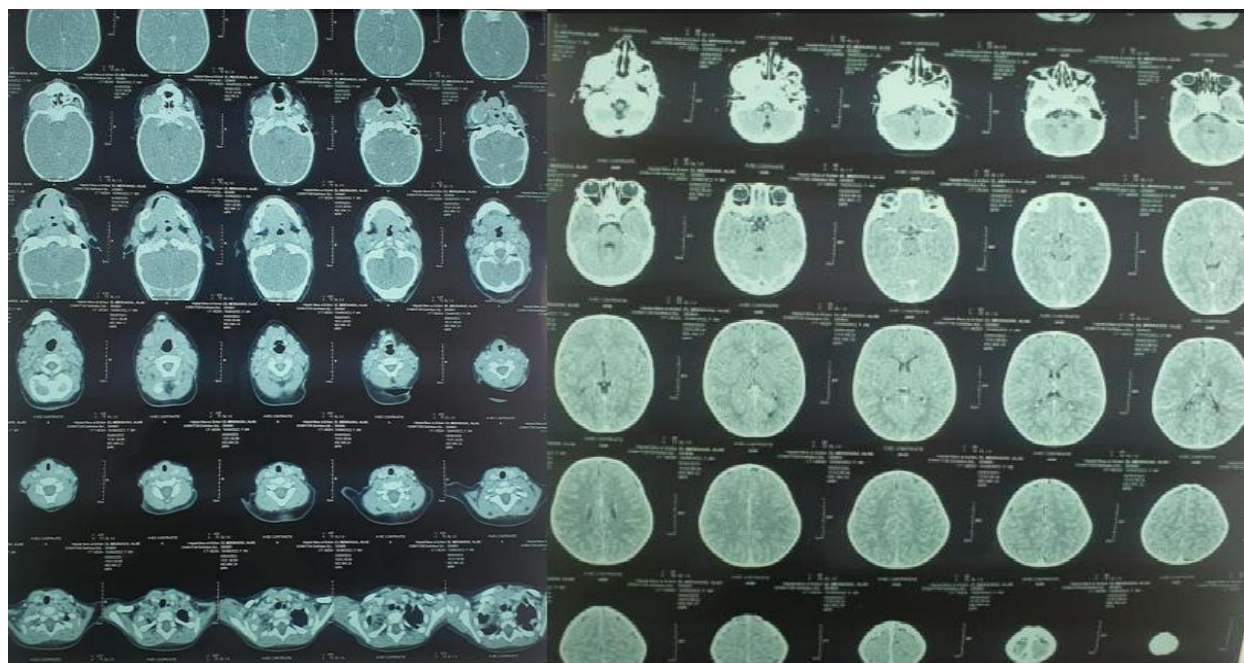
aspect, and displaces the premolars and molars; extends to the lateral pterygoid plate, causing focal bone lysis; infiltrates the pterygoid muscles, muscles of mastication, and temporalis muscle; bulges into the right lateral pharyngeal space and abuts the Eustachian tube, with complete opacification of the middle ear; abuts the right side of the tongue.

Laterally:

Displaces and thins the ascending ramus of the mandible, causing focal bone lysis, while sparing the temporomandibular joint.

Multiple lateral cervical lymphadenopathies; some have a short-axis diameter exceeding 1 cm [e.g., 18 x 14 mm in the right jugulocarotid chain].

Multiple bilateral soft-tissue nodules within the occipital muscles [largest measuring 7 x 5 mm]; ultrasound follow-up identifies these as lymph nodes.



The treatment consisted of the excision of the tumor mass. Under general anesthesia with the patient intubated, exploration revealed a mass adherent to the

right infratemporal fossa, and the tumor volume was reduced.

The operative procedure lasted 45 minutes.



Postoperative course [Day 1]:

Mild right cheek edema; no hematoma; clean surgical site.

Histopathological examination of the surgical specimen reveals an encapsulated, firm, multilobulated nodular mass weighing 25 g and measuring 5 x 3.2 x 5 cm. On sectioning, it appears beige-white with a fasciculated appearance.

Histological examination shows the nodular mass to be an encapsulated spindle-cell tumor

proliferation composed of elongated, monomorphic, spindle-shaped tumor cells with pale cytoplasm and nuclei lacking atypia or mitoses. The stroma is fibrohyaline, with focal myxoid areas.

Immunohistochemical analysis shows that the tumor cells express beta-catenin and—focally—SMA [smooth muscle actin], yielding a histological and immunohistochemical profile consistent with a desmoid tumor.

The postoperative course was marked by the recurrence, seven months after the procedure, of a right-sided mass involving the cheek and lateral neck. The mass progressively increased in size, extending from the

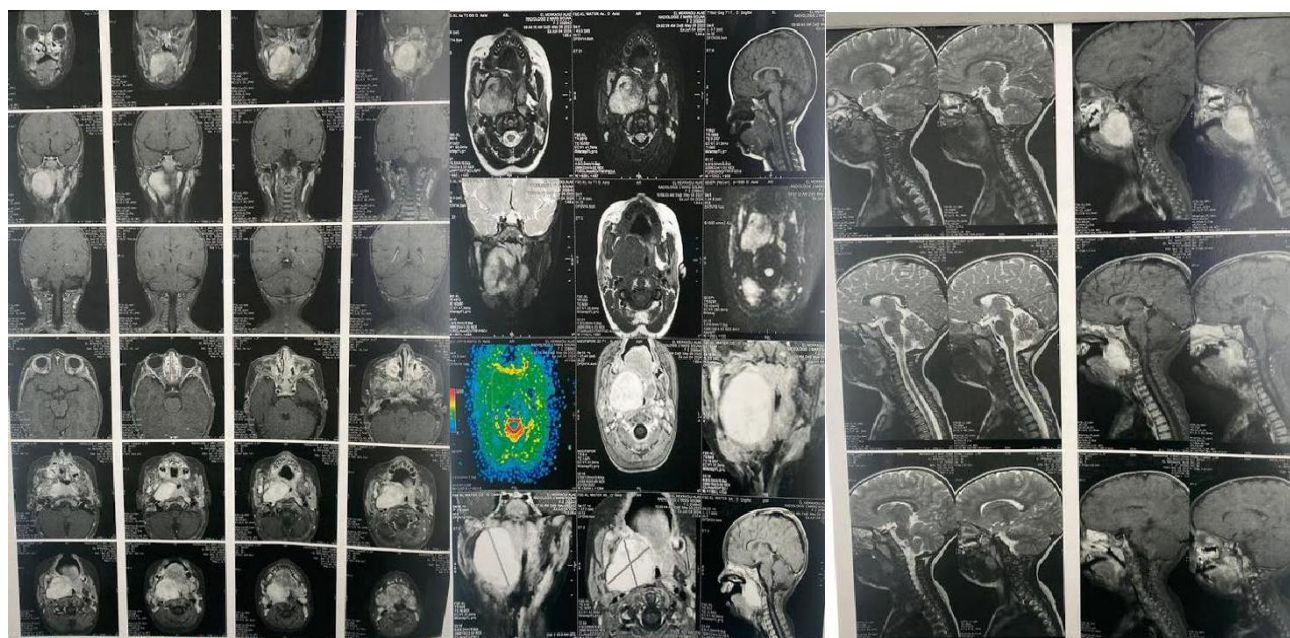
retroauricular region to the suprasternal notch; it was firm and fixed to deep structures but mobile relative to the superficial tissues, with no overlying signs of inflammation or infection.



MRI revealed a large mass involving the tonsillar region and the parapharyngeal space. It exhibits heterogeneous T2-weighted fat-saturated hyperintensity, mildly heterogeneous T1-weighted hypointensity, and mild diffusion restriction with intermediate ADC values; it shows strong, mildly heterogeneous contrast enhancement. It measures 42.3 mm in height, 43 mm in transverse diameter, and 33 mm in anteroposterior diameter. It displaces the right pterygoid muscles. It bulges into the oropharynx and extends to the tonsillar

fossa, causing its displacement. It abuts the deep lobe of the parotid gland, displacing it, and comes into contact with the internal carotid artery, displacing the vessel while leaving it patent.

It bulges into the right choana. The appearance is consistent with a residual mass in the deep spaces of the right face, extending from the nasopharynx to the oropharynx and displacing adjacent structures without significant invasion.



DISCUSSION

Desmoid tumors were first described in the abdominal wall by MacFarlane in 1832. The extra-abdominal variant of desmoid tumors was identified as a distinct entity by Nichols in 1923 [11].

They account for less than 0.03% of all tumors [12] and approximately 3.5% of fibrous tumors [13].

Infantile fibromatosis is the pediatric equivalent of fibromatosis [14]. This lesion presents between birth and age 8, most often during the first two years of life.

It is a histologically benign tumor with no metastatic potential, yet it exhibits a high propensity for local invasion and recurrence [5]. This leads to significant morbidity and even, at times, death, particularly in cases involving the head and neck.

Furthermore, it has been shown that these tumors are more aggressive in the pediatric population [15].

According to the World Health Organization classification of head and neck tumors, the desmoid tumor is defined as a borderline soft-tissue tumor with low malignant potential. It is characterized by local aggressiveness and a local recurrence rate of approximately 20%, but without metastasis. Of all desmoid tumor cases, 7% to 15% occur in the head and neck region [16].

Several theories regarding the etiopathogenesis of this condition have been proposed. The hormonal theory emerged following the discovery of estrogen and progesterone receptors on tumor cells. Viral and genetic theories have also been discussed, but the true origin of this pathology remains to be determined [17].

Clinically, the lesion generally presents as a recently appearing, poorly defined, slow-growing mass that is minimally tender or non-tender [18]. The development of such a lesion in the cervicofacial region—an area characterized by numerous functional structures and structures essential to aerodigestive functions—can lead to functional deficits [vision, chewing, swallowing, etc.] [19-20], nerve compression [21, 22], or even life-threatening complications.

CT scanning and, above all, magnetic resonance imaging [MRI] are the radiological examinations of choice. They allow for the assessment of tumor boundaries and its relationship with the neurovascular bundle [23].

Histological examination confirms the diagnosis by revealing a highly cellular appearance, consisting of fibroblasts and myofibroblasts organized into very long fascicles. Nuclear size varies moderately, and there are no signs of atypia or malignancy [24]; this rules out low-grade fibrosarcoma, reactive fibroblastic proliferation, and nodular fasciitis. The differential diagnosis also includes rhabdomyosarcoma, malignant histiocytoma, neurofibroma, and mesenchymal tumors [25].

Surgery is traditionally considered the best treatment and should be performed as soon as possible [at a stage where the tumor is completely resectable without excessive functional or cosmetic sequelae] [26]. The tendency of desmoid tumors to spread insidiously along muscles and fasciae—without forming a pseudocapsule or clearly defined boundaries—makes determining resection margins very difficult. Radical resection with wide surgical margins is therefore considered necessary.

However, a high rate of local recurrence is observed. The primary risk factor for recurrence is incomplete surgical resection with positive surgical

margins [R1-R2]. Other risk factors include young age, very large tumors, and specific tumor sites: head and neck tumors are associated with the poorest prognosis [27].

External beam radiotherapy is frequently used as an adjuvant treatment in adults, with well-documented efficacy [10, 12]. A recent meta-analysis showed that radiotherapy—whether used alone or as an adjuvant following surgery—resulted in better local control rates than surgery alone [18]. However, it can cause adverse effects [fibrosis, paresthesia, edema, fractures, skin ulcers] and potentially fatal radiation-induced secondary cancers [12, 18].

External beam radiotherapy exposes children to a considerable risk of long-term cosmetic and functional sequelae [radiation-induced inhibition of craniofacial bone growth, mandibular and facial bone hypoplasia, panhypopituitarism, scoliosis, hypothyroidism, neuropathic ulcers, osteomyelitis, and dermatitis with sometimes fatal chronic ulcers] [5, 6, 8, 11, 17, 19]. Radiotherapy is also associated with a risk of radiation-induced cancer within the treatment fields [basal cell carcinoma, papillary thyroid carcinoma, mandibular osteosarcoma] [5, 11, 18].

Desmoid tumors in children may also recur despite radiotherapy [8, 17]. In a series from St. Jude Children's Research Hospital, 11 out of 13 children relapsed after radiotherapy and 3 died from their disease [19], suggesting that radiotherapy for desmoid tumors in children does not provide local control as effectively as it does in adults.

Given these circumstances, radiotherapy should be avoided in children due to the high risk of sequelae and radiation-induced cancers, as well as its lower efficacy compared to adults.

Various cytotoxic and non-cytotoxic drug therapies have been proposed for the treatment of desmoid tumors. Non-cytotoxic medications include hormonal therapy, non-steroidal anti-inflammatory drugs, interferon-alpha, and imatinib. Hormonal treatment primarily consists of anti-estrogens such as tamoxifen [Nolvadex] and, more recently, toremifene—a promising drug that has yielded a partial response in 25% of cases, stable disease in 65%, and symptom relief in 75%, even following the failure of other hormonal treatments²⁰. Imatinib is a tyrosine kinase receptor inhibitor that has demonstrated efficacy in desmoid tumors. A phase II clinical trial reported a two-year progression-free survival rate of 55% with imatinib²¹. Another trial reported one-year progression-free survival with an objective response in 6% of cases²².

The efficacy of cytotoxic agents against desmoid tumors has been reported in numerous retrospective studies. Partial or complete responses have

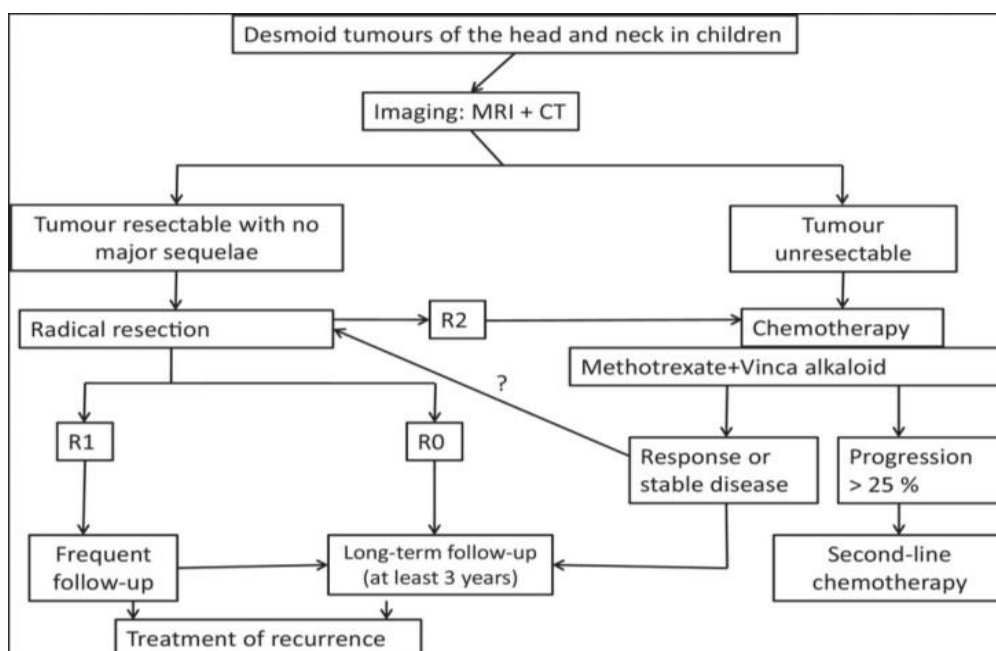
been observed with the VAC regimen [vincristine, actinomycin D, and cyclophosphamide], hydroxyurea, and the combination used in our study [methotrexate plus a vinca alkaloid] [6, 23]. In a retrospective study of 59 pediatric desmoid tumors, the overall response rate to systemic therapy was 33% [7]. No long-term adverse effects from chemotherapy were reported [8].

A phase II study demonstrated that methotrexate combined with vinblastine, administered weekly for several months, induced a partial response [40%] or prolonged stable disease [60%] in patients with unresectable desmoid tumors [24]. This chemotherapy regimen was also evaluated in a pediatric population, showing a response rate of 48% [7]. The Pediatric Oncology Group [POG] study 9650—evaluating the combination of methotrexate and vinblastine in patients under 19 years of age with recurrent desmoid tumors or tumors unsuitable for surgery or radiotherapy [25]—showed that this regimen was well-tolerated and potentially effective in children: a measurable response [complete, partial, or minor] was observed in 31% of cases, and stable disease in 38%. The most frequently reported toxicities were neutropenia, anemia, nausea, vomiting, and elevated transaminases; these were reversible upon discontinuation of chemotherapy. This

chemotherapy regimen appears suitable for treating this benign tumor, given the absence of long-term toxicity [25].

For patients who do not tolerate vinblastine, it can be replaced by another vinca alkaloid: vinorelbine [8]. The combination of vinorelbine and methotrexate—included in the 2005 European Paediatric Soft Tissue Sarcoma Study Group [EpSSG] guidelines for Non-Rhabdomyosarcoma Soft Tissue Sarcoma [NRSTS]—appears to be as effective as, yet less neurotoxic than, the vinblastine and methotrexate combination [13]. Marked tumor progression [$\geq 25\%$] in response to chemotherapy should be considered a treatment failure, warranting the consideration of second-line therapy [13].

However, a "wait-and-see" approach does not appear justified in children, given the more aggressive nature of desmoid tumors in this population and the particularly high morbidity and mortality associated with head and neck tumors. Furthermore, the risk of progressive disease appears to exceed the rate of spontaneous regression. In light of all these data, a management algorithm for pediatric head and neck desmoid tumors can be proposed.



Excision must be complete and wide, adhering to strict oncological surgical principles to minimize the risk of recurrence. The risks associated with radiotherapy—specifically impaired facial growth, soft-tissue atrophy, hypopituitarism, and radiation-induced cancers—limit its use to inoperable tumors and cases of incomplete resection [2]. Chemotherapy may be used alone or preoperatively to reduce tumor size. Several protocols have been proposed; the most effective is the VAC regimen, combining vincristine, actinomycin, and cyclophosphamide [4]. Other therapeutic approaches are

currently being evaluated and appear to be effective alternatives: interferon-alpha, 1,25-dihydroxyvitamin D3, anti-estrogens such as tamoxifen, and non-steroidal anti-inflammatory drugs [4].

CONCLUSION

Desmoid tumors account for less than 0.03% of all tumors [3] and approximately 3.5% of fibrous tumors [4]. Surgery currently remains the standard treatment, despite a high recurrence rate reaching 70%. Excision must be complete and wide—adhering to strict

oncological surgical principles with negative margins—to minimize the risk of recurrence. Chemotherapy may be proposed to reduce tumor size [5]. Long-term follow-up is required given the risk of recurrence [6].

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