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Chapter
Soft-Tissue Tumors of the Head and Neck Region

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Abstract

Fibroblastic and myofibroblastic neoplasms in the head and neck region are a rare group of tumors ranging from benign lesions to malignant lesions. Due to the difficult anatomy of the head and neck region, even neoplasms without metastatic potential can pose significant therapeutic challenges in this region. In this section, the most common soft-tissue neoplasms in the head and neck region will be discussed.

Keywords: head, neck, soft tissue

1. Introduction

A general approach to fibroblastic and myofibroblastic neoplasms is presented in this section. Adequate clinical information and biopsy material are required to diagnose soft-tissue tumors. The patient’s age, tumor location, growth characteristic, and rate of growth play an important role in diagnosis. There are significant differences between soft-tissue tumors seen in children and those seen in adults. The location of the tumor is important in terms of differential diagnosis. Sarcomas are usually located deep, but benign and low-grade tumors are located superficially [1].

2. Pathological diagnosis of soft-tissue tumors

Diagnostic biopsies for soft-tissue tumors can be performed differently, depending on the location. Excisional biopsy may be preferred for superficial masses and size suitable for complete removal. Incisional biopsy or FNAB (Fine needle aspiration biopsy) may also be preferred for large masses [2].

3. Radiological imaging of soft-tissue sarcoma

3.1 Direct X-ray

Direct X-ray can be helpful in the diagnosis of soft-tissue tumors, with important findings such as calcification in the mass or detection of invasion in the adjacent bone. Calcification can be seen in many benign tumors, such as synovial sarcoma and epithelioid sarcoma.
3.2 USG (ultrasonography)

Ultrasound is an easily accessible and reproducible method and is the first method of evaluation mostly for superficial soft-tissue tumors. Ultrasound is a limited method in deep and large lesions. Ultrasound is the fastest informative method in distinguishing the internal structure of the lesion from cystic/solid. It can also offer biopsy with imaging. The vascularity of the tumoral lesion can be evaluated with Doppler USG.

3.3 Magnetic resonance imaging (MRI)

Magnetic resonance imaging provides important information in diagnosis, staging, and treatment follow-up with its multiplanar imaging capability and superior soft-tissue resolution [3].

3.4 Diagnosis and determining of treatment response in soft-tissue sarcoma with PET/CT

Soft-tissue sarcomas are rare mesenchymal cell tumors that arise from connective tissue, fat, muscle, vascular, and nerve tissues. F-18 FDG PET/CT can be used for initial diagnosis, staging, grading, therapy monitoring, and radiotherapy planning. PET-CT can also be used to evaluate changes that occur after recurrence and radiotherapy. F-18 FDG PET/CT cannot differentiate between benign or malignant sarcomas, but it may help distinguish between high and low-grade malignant sarcomas [4].

4. Nodular and cranial fasciitis

Cranial fasciitis is a fibroproliferative lesion of the scalp most commonly seen in the very young pediatric population. Cranial fasciitis and nodular fasciitis are histologically similar. The differences between these two lesions are the location and patient age [5]. Nodular fasciitis (NOF) is a benign myofibroblastic neoplasm that presents as a solitary subcutaneous mass on the upper extremities, trunk, or head and neck. In the head and neck, NOF most commonly arises on the face or neck but can also be seen in the oral cavity, orbit, parotid, and ear [6]. Although the age range at presentation is wide, the peak incidence is in the third and fourth decades. Tumors grow rapidly, typically in less than 3 months, may be painful or painless, and are generally lesions less than 3 cm. Spontaneous regression before surgical resection is characteristic, with “recurrences” only occurring after incomplete surgical excision [7]. Cranial fasciitis (CF) is a rare variant of NOF that arises on the scalp, most commonly in the temporal and parietal regions. In contrast to NOF, CF typically presents in infants less than 2 years of age, including some congenital tumors. Lesions often cause erosion of the outer table of the skull but occasionally erode through the inner table as well. On examination, NOF and CF are rubbery, fibrous, or myxoid, focally cystic masses, which can appear circumscribed or some infiltrative [8]. Although there is no definitive evidence, trauma can be shown in the etiology of both CF and NOF [9]. Because these tumors are self-limiting, the concept of transient neoplasia has been proposed [10]. Treatment for cranial fasciitis is surgical resection [11].
5. Fibrous hamartoma of infancy

Fibrous hamartoma of infancy (FHI) is a rare benign neoplasm that presents as a painless, solitary, subcutaneous mass in the axilla, trunk, or proximal extremities. A total of 10% of cases occur in the head and neck region, including the cheek and scalp [12]. FHI is typically seen under 2 years of age, is more common in male infants, and occurs congenitally in 20% of cases [12]. Local recurrence can be seen in 15% of FHI cases without invasion and metastasis [12]. FHIs are lesions ranging from 3 to 5 cm, containing varying amounts of fat and fibrous tissue. FSI histologically includes three components as spindle cell fascicles, mature adipose tissue, and nodules of primitive mesenchyme composed of spindled to stellate cells within a loose basophilic or myxoid stroma [13]. FHI treatment is local excision [14].

6. Nasopharyngeal Angiofibroma

Nasopharyngeal angiofibroma (NA), which occurs most commonly in adolescent males, is a rare fibrovascular neoplasm of the posterolateral nasal wall [15]. NA classically has three clinical manifestations—nasal obstruction, recurrent epistaxis, and nasopharyngeal mass. NA tumor can be locally aggressive, sometimes it can spread to the paranasal sinus, skull base, and intracranial [15]. NA has been shown to be associated with familial adenomatous polyposis in some cases [16]. The etiology of NA is unknown, but its occurrence in adolescent males suggests that hormonal factors are important [17]. Imaging methods are sufficient for diagnosis. There is no need for a biopsy to make a preoperative diagnosis. Postoperative recurrence can be seen in a quarter of cases [18]. Macroscopically, NA is typically a polypoid or lobulated lesion [19]. NA histologically consists of multiple vascular spaces of variable size within a fibrous stroma containing plump spindle to stellate stromal cells [20]. The androgen receptor is located in stromal cells [21]. NA shows high positivity in nuclear staining for β-catenin [22]. The testosterone receptor blocker flutamide can be used in the treatment of stage I and II tumors [23]. Conformal radiotherapy provides a good alternative to conventional radiotherapy in advanced diseases, such as diffuse juvenile nasopharyngeal angiofibroma (JNA) or intracranial spread [24]. Biopsy is contraindicated in JNA. Surgically, a lateral rhinotomy, transpalatal, transmaxillary, or sphenoethmoidal route is used for small tumors. The infratemporal fossa approach is used when the tumor has a large lateral extension. The midfacial degloving approach can be used for improves posterior access to the tumor [25].

7. Nuchal-type and Gardner fibromas

Nuchal-type fibroma (NTF) and Gardner fibroma (GAF) are two histologically similar yet distinct benign fibroblastic tumors that arise within different age groups and at different body sites, allowing for distinction in most cases. GAF, which affects males and females equally. NTF has a strong male predominance. NTF occurs most commonly in the 5th decade of life, while GAF is more common in young children, although age ranges are wide for both tumor types. While NTF mostly occurs in the back of the neck, it can also occur in the nape areas, including the face and upper back [26]. GAF mostly occurs in the trunk and paraspinal area, only 15% is seen in the
Advances in Soft Tissue Tumors

head and neck region. Desmoid fibromatosis occurs largely in the same region as GAF [27]. The vast majority of GAF are associated with FAP and APC germline mutations [28]. Simple excision is curative. Patients can develop a recurrence, so follow-up is required [29].

8. Desmoid fibromatosis

Desmoid fibromatosis (DF) is a locally aggressive fibroblastic neoplasm, which occurs in the head and neck region in about 15% of cases. It occurs in the head and neck region in the majority of pediatric cases. Although structures such as the face, mandible, paranasal sinuses, and larynx are affected, the neck is most affected in the head–neck region [30]. As it can be seen at any age, it is most common in childhood and young adults. Clinically, a painless and rapidly growing mass is seen. However, sometimes pain and neurological deficits can be seen [31]. DF occurs in the head and neck region, as in other parts of the body, as a result of trauma or, rarely, APC germline mutations. DF in the head and neck region has high morbidity due to its therapeutic challenges, unpredictable tumoral behavior, and proximity to vital anatomical structures. DF left untreated is generally stable and even spontaneous regression may occur. It can be locally aggressive, with a recurrence rate of up to 30% after surgical excision. Treatment is typically surgery, but in recent years conservative approaches such as radiotherapy and chemotherapy have been preferred [31]. Macroscopically, DF appears white-tan, whorled, and fibrous, with ill-defined borders. Histologically, the lesions are comprised of bland spindled to stellate fibroblasts arranged in long sweeping fascicles within a collagenous stroma, irregularly infiltrating through surrounding adipose tissue or skeletal muscle [32]. Somatic mutations in exon 3 of CTNNB1 are found in up to 90% of sporadic DF [33].

9. Dermatofibrosarcoma protuberans and giant cell fibroblastoma

Dermatofibrosarcoma protuberans (DFSP) and giant cell fibroblastoma (GCF) are two cutaneous fibroblastic neoplasms that share clinicopathologic and genetic features. GCF predominantly affects pediatric patients, but DFSP arises most often in young to middle-aged adults, although both tumors can affect newborns to elderly individuals. Both seen males predominantly and present with a slow-growing, painless, often protuberant, multinodular, or polypoid cutaneous mass or plaque. While both occur most frequently in the trunk and proximal extremities, GCF is rarely seen in the head and neck region, while DFSP is seen in 15% of cases [34]. Local recurrence is seen in 50% of cases in cases of GCF and DFSP, inadequate resection. GCF and DFSP do not metastasize, but the fibrosarcomatous variant of DFSP typically metastasizes to the lungs in 15% [35]. Excision with wide surgical margins is the treatment of choice. Tyrosine kinase inhibitors can be used in conservative treatment. Generally, DFSP and GCF are infiltrative, predominantly dermal and subcutaneous lesions, but DFSP arising on the scalp may invade the periosteum or the skull. Histologically, both neoplasms display honeycomb infiltration through subcutaneous fat, often sparing entrapped skin adnexal structures [36]. The standard treatment of DFSP is surgical excision at all stages. Initial resected tumors with positive margins or relapsed/recurrent tumors need to be additionally resected [37]. Adjuvant
radiotherapy is used to reduce the incidence of both local recurrence and metastasis [38]. The typical treatment for giant cell fibroblastoma is surgical resection. Surgery should be performed as wide and complete tumor resection [39].

10. Solitary fibrous tumor

Solitary fibrous tumors (SFT) are more common in middle age and are equally common in both sexes. SFT can see in any part of the body. It is seen in the sinonasal, orbit, oral cavity, and salivary gland regions in the head and neck region proportion of %10–15. Head and neck SFTs typically present as slowly growing painless masses [40]. Although the local recurrence rate is high in head and neck SFTs, distant metastasis rates are low. Recurrences may occur greater than 15 years after primary excision. Advanced age, tumor size, increased mitotic activity, and tumor necrosis are among the factors that increase the risk of metastasis and death [41]. Generally, head and neck PFTs are limited, solid, white-tan, and fibrous lesions that may show infiltrative growth and sometimes bone invasion. Histologically, SFTs are morphologically heterogeneous [42]. Rarely, high-grade sarcomatous transformation may be seen [43]. Characteristically, SFTs are diffusely positive for CD34 [44]. The treatment of both benign and malignant SFT is complete en bloc surgical resection [45].

11. Inflammatory myofibroblastic tumor

The inflammatory myofibroblastic tumor is a myofibroblastic neoplasm that occurs in the lungs, abdomen, and pelvis [46]. About 15% occur in the head and neck, where they are more common in adults. Laryngeal IMTs present with hoarseness and dysphonia while sinonasal IMTs typically present with nasal obstruction and pain [47]. While up to 30% of IMTs have systemic symptoms and laboratory abnormalities, this is not seen in sinonasal or laryngeal IMTs [48]. Laryngeal IMTs generally arise in the glottis and follow a benign clinical course following excision while sinonasal and oral cavity IMTs are clinically more aggressive, with higher rates of recurrence, metastasis, and mortality despite treatment [49]. Roughly, IMTs appear polypoid or nodular, typically less than 3 cm in the larynx and less than 7 cm on the head and neck. Histologically, IMTs may show a myxoid fasciitis-like pattern, a cellular spindle cell pattern, and a hypocellular fibromatosis-like pattern, and sometimes all can be seen within the same lesion [50]. The recommended treatment for IMT is total resection of all tumor tissues [51].
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