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Case Report

Fasciitis Ossificans of the Larynx

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ABSTRACT

We describe a rare case of laryngeal fasciitis ossificans. A 58-year-old man presented with hoarseness and a nodule was found in the larynx. Excisional biopsy was performed, and follow-up laryngoscopy showed complete resolution of this reactive lesion, and normal laryngeal function. The 0.6 cm diameter nodule was well circumscribed and histologically, the lesion was composed of uniform woven bone trabeculae with rimming of osteoblasts and cellular stroma. At the periphery, uniform spindle cells actively proliferated in edematous stroma. Spindle cells were immunoreactive for vimentin and α-smooth muscle actin, suggesting myofibroblastic differentiation. Fasciitis ossificans is histologically identical to myositis ossificans, but tends to present no zonation phenomenon. Fasciitis ossificans is a rare form of heterotopic bone formation, commonly presenting with signs of local inflammation or pain. This patient's successful outcome suggests that conservative resection may be both diagnostic and curative.

Keywords: Myositis ossificans, Larynx, Fasciitis

Introduction

Fasciitis ossificans is a rare subtype of nodular fasciitis, a rapidly growing benign reactive lesion, and was first described by Kwittken and Branche in 1969 (1). Nodular fasciitis commonly occurs in the forearm and arm, but the laryngeal presentation is extremely rare, and only about 15 cases have been described in the literature (2, 3).

Fasciitis ossificans is histologically identical to myositis ossificans, an intramuscular reactive lesion with ossification (4). However, the zonal pattern, which is characteristic for myositis ossificans, presenting a fibro-edematous center that is histologically the same as nodular fasciitis, trabecular osteoid in the middle zone, and mature bone at the periphery, is usually lacking (3,5). The most frequently affected site by inflammatory

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myofibroblastic pseudotumour (IMFPT) is the lung; a much less common location is the larynx.The lesion progressed uniquely to show myositis ossificans-like maturation over time (6).

Myositis ossificans progressiva is a rare disease, histologically similar, but with autosomal dominant pattern of inheritance leading to complete ossification of the muscular system and is usually associated with anomalies of the hands and feet (7).

Another similar lesion at light microscopy, heterotopic ossification, the formation of bone in soft tissue, requires inductive signaling pathways, however, little is known about the molecular pathogenesis of this condition (8). Also, fasciitis ossificans may mimic extraskeletal and parosteal osteosarcoma, radiographically, clinically, and/or histologically. Thus, it is important to clearly distinguish this lesion from a malignant process (4,5). We report a case of this uncommon laryngeal lesion.

Case report

A 58-year-old man with a past history of hoarseness, presented with a nodule in the larynx. The hoarseness had been present for 3 months. The patient had no distinctive past medical or family history.

On direct laryngoscopic examination, a 0.6 cm ×0.5 cm polypoid lesion on the left anterior side of glot was noted. An excisional biopsy was performed and all the symptoms resolved on follow-up the patient. The biopsy specimen was fixed in 10% neutral-buffered formalin and decalcified. Tissue sections of 4 μm in thickness were cut for hematoxylin and eosin staining and immunohistochemical studies. The primary antibodies used were vimentin (NP018, Dako,1/30), α-smooth muscle actin (NP025, Dako, 1/50), pancytokeratin (NP046,AE1/AE3, Dako, 1/100), EMA(NP022,Dako,1/100), desmin (NP041, Dako, 1/100), S-100 protein (Dako ,1/50), c-kit (polyclonal, M7140, Dako, 1/100). Macroscopically, the nodule was well-circumscribed and 0.6 cm in diameter. Microscopically, the nodule was composed of thick woven bone trabeculae and fibrous stroma. The woven bone trabeculae were essentially uniform, broad, hypercellular, and rimmed by osteoblasts .In the periphery of the lesion, spindle cells actively proliferated in edematous stroma, and reactive osteoid was recognized (Fig.1).We counted 3 mitotic figures per 10 high-power fields, but observed no atypical forms. Immunohistochemically, spindle cells were positive for vimentin and α-smooth muscle actin (Fig. 2) but negative for pancytokeratin (AE1/AE3), EMA, desmin, S-100 protein, or c-kit, suggesting myofibroblastic characters.

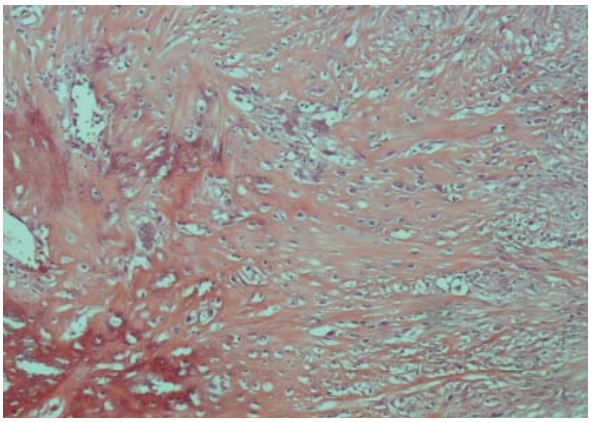

Fig. 1- Woven bone trabeculae are regular, broad, hypercellular, and rimmed by osteoblasts and occasional osteoclasts (H&E staining×100)

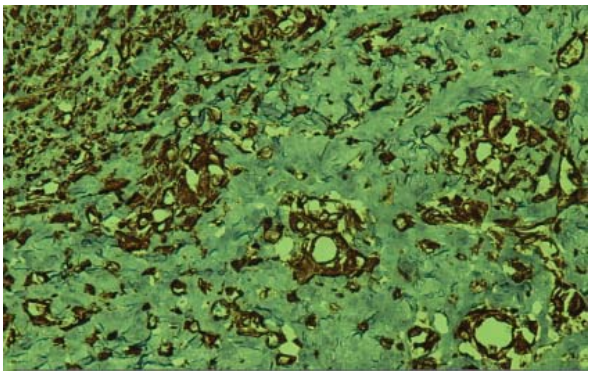

Fig. 2- Spindle cells were positive for vimentin

Discussion

Metaplastic bone formation of the larynx is a rare occurrence in inflammatory diseases as well as in malignant or benign tumors. The centrally located trabecular bone and proliferating spindle cells at the periphery seen in this case are noted to have reverse zonation pattern, generally implying bone-forming sarcoma (4).

The zonation phenomenon is characteristic of myositis ossificans, a histologic feature the same as for nodular fasciitis in the central portion, trabecular osteoid in the middle zone, and mature bone in the outer layers (4). Fasciitis ossificans is a benign process of abnormal, extraskeletal ossification occurring in inflamed fascial tissue, in response to soft tissue trauma, surgery or without any known precipitating factor (9). Proposed mechanisms for atraumatic fasciitis ossificans include unreported trauma, repeated minor mechanical injuries, and local ischemia or inflammation (10).

In this case, the nodule was distinguished from osteosarcoma because the spindle cells presented uniform nuclei without increased chromatin, pleomorphism, or evident nucleoli. There was no atypical mitotic figure, and the woven bone trabeculae were regular. Also, fasciitis ossificans may mimic extraskeletal and parosteal osteosarcoma, radiographically, clinically, and/or histologically. It is of paramount importance to clearly distinguish this lesion from a malignant process. Extraskeletal osteosarcoma,comprising less than 5% of all osteosarcomas, typically effects an older population and most commonly arises in the thigh. Histologically, it shows areas of necrosis and atypical nuclei and may show a "reverse zoning phenomenon". There is frequently infiltration of adjacent soft tissues (4, 5, 11).

Fasciitis ossificans is a bone-forming subtype of nodular fasciitis, and, histologically, a closely similar lesion to myositis ossificans, parosteal fasciitis, panniculitis ossificans, and fibro-osseous pseudotumor of the digits. Heterotopic bone formation may occur in any soft tissue including muscle (myositis ossificans), nerve (neuritis ossificans) or fascia (fasciitis ossificans) (12). Heterotopic ossification, the formation of bone in soft tissue, requires inductive signaling pathways, inducible osteoprogenitor cells, and a heterotopic environment conducive to osteogenesis. Little is known about the molecular pathogenesis of this condition. Research into two rare heritable and developmental forms, fibrodysplasia ossificans progressiva and progressive osseous heteroplasia, has provided clinical, pathologic, and genetic insights (8).

Myositis ossificans develops in the muscles which vary in histologic features with different amounts of immature fibroblasts, osteoid, cartilage, and mature bone (13). This disease is felt to have an autosomal dominant pattern of inheritance and is usually associated with anomalies of the hands and feet. Afflicted patients are frequently misdiagnosed in childhood as having a rheumatologic disorder. The correct diagnosis becomes clear, later in life when true bone is formed in striated muscle, ligaments, and fascia. Although muscles of the heart, diaphragm, larynx, and sphincters are spared, those of the chest wall are not, and pulmonary function is progressively deteriorates (7, 13).

The present nodule histologically corresponds to type-II lesion of myositis ossificans is classified by Sumiyoshi *et al.* (13), which mainly consists of osteoid and immature trabecular bone rimmed by osteoblasts intervened by loose fibrous connective tissue. Parosteal fasciitis, panniculitis ossificans, and fibro-osseous pseudotumor of the digits are the terms used when the corresponding le-

sions arise in the parosteal region, subcutaneous fat tissue, and digits, respectively (4). The same lesions are named as fasciitis ossificans when they occur elsewhere in the soft tissues away from the periosteum (4).

Fasciitis ossificans may be differentiated from inflammatory myofibroblastic pseudotumour (IMFPT) a rare condition most frequently affects lung; much less common, larynx and subglottis. Previous trauma is purportedly one potential aetiological factor. Furthermore, the lesion progressed uniquely to show myositis ossificans-like maturation over time (6).

Several reports have described heterotopic ossification presenting with a peripheral neuropathy (14), including two reported cases of myositis ossificans presenting with a radial nerve neuropathy (15, 16); and one case of fasciitis ossificans presenting as a peripheral neuropathy. In the breast, to the best of our knowledge, only one case of fasciitis ossificans has been described (2). We considered the present lesion as fasciitis ossificans because it was situated at the superficial layer of the larynx with signs of local inflammation and pain.

The present nodule was unusually composed of mainly thick woven bone trabeculae and had a minor component of nodular fasciitis-like hypercellular edematous stroma and reactive osteoid. The early phase of fasciitis ossificans is presumed to be similar to nodular fasciitis and thereafter osteoid formation progresses (4).

In nodular fasciitis, chromosomal translocation involving chromosome 15 and immunohistochemical overexpression of TRKC tyrosine protein kinase have been shown (17). Cytogenetic abnormalities in fasciitis ossificans or myositis ossificans have not been reported, and it is not evident whether a common pathway is associated with their development. The mechanisms of osteoid formation in fasciitis ossificans are also not fully elucidated, similar to the overall metaplastic ossification process.

Simple excision is recommended and is sufficient for a definite histological diagnosis and therapy because neither fasciitis ossificans nor nodular fasciitis generally recur. There are cases described with spontaneous regression following incomplete excision or only fine needle aspiration (18). No transformation to sarcoma has been reported (10, 18). To avoid an unnecessarily aggressive treatment, in the differential diagnosis for larynx lesions, fasciitis ossificans, a benign bone-forming nodule, needs to be recognized as a bony hard tumor that can develop in the larynx.

Acknowledgements

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