FIBROMATOSIS OF PAROTID GLAND: A RARE LOCALIZATION

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ABSTRACT

Desmoid tumors are rare, histologically benign but local aggressive tumors that belong to the group of the deep fibromatosis. They are usually seen in abdominal region but rarely could be located in the head and neck. We report a case who was operated with the diagnosis of fibro-matosis and discuss the etiology, clinical presentation, pathology and various treatment mo-dalities.

***Key words**: Desmoid tumor, head and neck, parotid gland.*

***Received** Septemper 18, 2013; **Accepted** October 24, 2013*

Introduction

Desmoid tumor that is also known as aggressive fibromatosis is a rarely seen benign tumor of parotid gland[1, 2, 3]. These tumors, originating from muscles and aponeurosis, are prolifera-tive diseases of fibrous tissue[2, 4, 5]. They are locally invasive and prone to recurrences but do not lead to distant metastasis. The incidence of extra abdominal fibromatosis in head and neck region is about 12%, of which the 3% originate in the parotid gland[6]. A case of rarely seen desmoid tumor of parotid gland is presented in this paper with its characteristic clinical and histopatological features.

Case presentation

A 56 year-old female patient presented complaining of a left parotidal mass, which has been growing for the last 6 months. A dense and fixed mass of 8x7 cm in size was detected in her left parotid lodge by physical examination. Her facial nerve functions were not influenced. Her blood levels were within normal limits (white blood cells (WBC) 6.79 x 10^3/uL [Normal Range (NR) 4.60 - 10.2 x 10^3/uL], hemoglobin (HGB) 13.9 g/dL [NR 12,2 - 18,1], platelets (PLT) 168 x 10^3/uL [NR 142 - 424 x 10^3/uL]. In her neck Magnetic Resonans (MR) imaging showed a lesion of 4x3x6 cm in size located deeply in left parotid gland that was hypointense in T1 sequences and showed low signal intensity in T2, and which also held frequent contrast matter in contrast surveys and lymph nodes in left submandibular. Fine nee-dle aspiration biopsy (FNAB) was reported as inflammatory/neoplastic proliferation leading to mesenchymal proliferation. The case was operated in our clinic and was applied left total parotidectomy. The solid mass invading surrounding tissues was extracted together with nor-mal surrounding tissues (Figure 1).

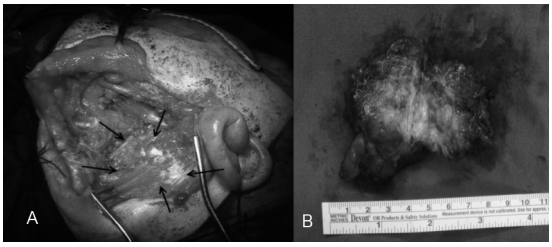

Figure 1: **A**) Intraoperative Picture shows the mass (arrows) **B**) The resected tumor (macros-copic apparence), the incised surface is white and fibrotic.

Pathological analysis of the mass resulted in fibromatosis (desmoid type). In the control MR images, which were performed in result of swelling and pain in the same location 18 months later, a well-bounded mass of 5x3x4 cm in size consistent with relapse was detected in left parotidal region (Figure 2 a).

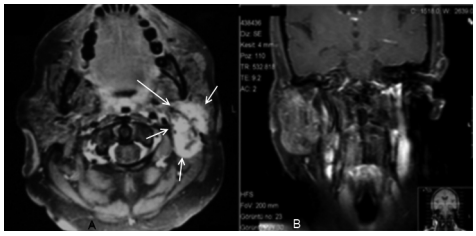

Figure 2: **A**) Axial MRI view shows the tumor originating from the left parotidal region (whi-te arrows), **B**) T1-weighted gadolinium-enhanced magnetic resonance imaging of the patient's neck shows that not distinguish in favor of recurrent contrast enhancement in the left parotidal region.

Therefore, the patient was re-operated. The fibrotic mass whose boundaries were not obvious, reaching sternocleidomastoid muscle in posterior, internal jugular vein in medial and mastoid apex in posterosuperior, was extracted together with surrounding tissues. Postoperative pathology report was "fibromatosis that stained focal cytoplasmic with actin; desmin, S-100 and cytokeratin negative and whose ki-67 proliferation risk is 1%" (figure 3 a-b). Since tumor persisted in surgical boundaries, adjuvant radiotherapy was applied. She is well with no evidence of recurrence approximately 4 years after surgery (figure 2 b).

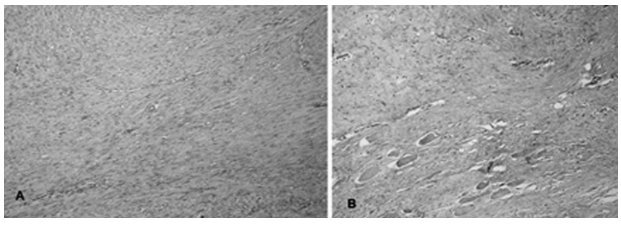

Figure 3: **A**) Histologic picture of the tumor displays spindle and collagen deposits typical of fibromatosis (HEX100). **B**) Fibromatosis that invaded muscle tissue (HEX100).

Discussion

Desmoid tumors are benign fibrous tumors that originate from musculoaponeurotic tissues (2, 7). Although local invasion and post-resection recurrence is frequent, no distant metastasis is observed. They do not have the potential for malignity (2, 8). Clinically, these tumors are between benign fibroma and well-differentiated fibrosarcomas and are intermediate-grade neoplasms as biological behavior[9, 10]. While several systems were proposed for categorization of fibromatosis, the most widely used is what Enzinder and Weiss described[2, 8]. Fibromatosis is mainly divided into two groups as superficial (originating from fascia) and deep (musculoaponeurotic) according to tissues they originate from. Superficial fibromatosis are small sized and they grow slowly and rarely advance into deep tissues. Deep ones, howev-er, are aggressive, grow faster, may reach larger size, and local relapse is seen in higher rates. Deep fibromatosis can be abdominal or extra-abdominal.

Desmoid tumors are relatively rare. Of all fibromatosis, only 12-15% is seen in head & neck region, and of head and neck fibromatosis, 40-85% are seen in neck[2, 3].

Although it is observed most commonly in cervical and supraclavicular region, among head and neck; cases of face, oral cavity, scalp, paranasal sinus and orbita are also seen[2, 6, 9, 11]. Of all the casesof head and neck fibromatosis just the 3% are located in parotid gland[7].

These tumors usually appear as painful, deeply located fixed mass, which grow slowly[10]. They may extend through facial plan and may lead to pain, respiratory distress, proptosis and epistaxis, invading surrounding tissues.

The pathogenesis of fibromatosis is not clear. Although several physical, endocrino-logical and genetic hypotheses are proposed, not a single mechanism could be discovered[2,5,12].

Trauma is the most important among physical factor. In addition blunt trauma and surgical incision are also held responsible. No specific cause among above-mentioned factors was de-tected in our case. It is more frequent in females and usually seen in third decade. Our case is elder than this age group. A palpable and painful mass is prominent in desmoid tumors. It is non-inflammatory and gives symptoms since they compress surrounding tissues.

A prominent radiological feature differentiating desmoid tumors from malignant soft tissue tumors is lacking[3,13]. MRI is more sensitive in detection of local relapse and soft tissue infiltrations. It is seen as multilobulated mass in heterogeneous density. In case gadolin-ium is administered, the tumor is seen as hypointense in T1 and varying from hypointense to hyperintense in T2[7].

Diagnosis of desmoid tumors requires histopatologic examination. Fine needle aspiration biopsy may not be sufficient because of desmoid tumor is relatively hypocellular as in our patient.

Differential diagnosis includes neurofibroma, reactive fibrosis, myxoma, nodular fasciitis, keloid, fibrous hamartoma and fibrosarcoma[2, 10, 12].

The best treatment for desmoid tumors is surgical resection with negative margine[2,5]. De-pending on its close relationship with vital organs, its aggressiveness and complexity of facial plans; it is difficult to extract head and neck fibromatosis without leaving negative surgical boundary. Hence, recurrence is more frequent in head and neck region compared to other re-gions[8, 9,12]. Although preoperative facial nerve functions of our case was normal, buccal and marginal branches of facial nerve covered by tumor had to be sacrificed in the second surgery.

Radiotherapy should be taken into consideration in location-related non-operable or partially extracted tumors. Radiotherapy may provide high local control[13]. Other treatments of choice include NSAID's (nonsteroidal anti inflammatory drugs), hormone therapy and chemotherapy[2, 3, 9, 14].

Consequently, although fibromatosis is benign histologically, its treatment is varying due to its aggressive clinical behavior and local infiltration leading up to relapse. It is an important morbidity cause in case it locates in head & neck region. Complete resection is the most effec-tive treatment of choice. In case surgery is either unsuccessful or ineligible, radiotherapy or chemotherapy should be taken into account. Close follow-up is crucial because of its high relapse incidence.

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