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Case Report

Subcutaneous dermatofibrosarcoma protuberans in parotid region: Case report

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Abstract. Dermatofibrosarcoma protuberans (DFSP) is a low-grade soft tissue sarcoma (fibrosarcoma) originated from dermal and subdermal layer of the skin. But subcutaneous localization is quite rare. It is one of the rare tumors and accounts for 0.1% of all malignancies. The tumor infiltrates into the deeps in the villous and finger shape. For this reason, it is quite difficult to get surgical margins in the tumor excision and the recurrence is a problem encountered frequently. An excision or Mohs technique of micrographic surgery is suggested by leaving a surgical space up to 5 cm from the tumor margines. In this case report, a case with DFSP atypically subcutaneous localized on superficial parotis gland was presented. It should be kept in mind that probability of the microscopic spread is high in these patients and they should be followed by reminding that the disease can recur in the following long years.

Key words: Subcutaneous, dermatofibrosarcoma protuberans (DFSP), parotid gland, and superficial parotidectomy

1. Introduction

Dermatofibrosarcoma protuberans (DFSP) is a locally aggressive soft tissue sarcoma originating from dermal and subdermal tissue of the skin (1). These tumors are characterized by the slowly progressive, firmly viscous on dermal endured plates, painless and red-brown lesions (2). Although it is frequently seen in the torso and at the proximal end of the extremity, it can also develop on any part of the body. It generally affects males between 20 and 50 years old (3). Although DFSP is a rare benign tumor, it has a high recurrence rate. This tumor can be rarely seen on head-neck region and defined subdermal localization (4,5). The reported case differs in that it imitates the parotid mass with the subcutaneous localization in parotid region without a dermal invasion.

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2. Case report

A fourty-seven year old male patient referred to our clinic with a mass in front of his right ear. The mass was firstly noticed five years ago and it got bigger and hardened within the elapsed time. In physical examination of the patient, the mass was solid, mobile and painless with palpation. The skin was in its natural appearance in the inspection. The chest radiography and routine laboratory values were normal. The mass was reported to be a well-circumscribed, heteregenous and hyperechoic lesion which has 5x2.5cm dimension on surface of parotid gland in the parotid ultrasonography (USG). Fine-needle aspiration biopsy (FNAB) was reported suspicious for malignancy with a small amount of chondromyxoid intermediate and pleomorphic atypical cells that have a broad vacuole cytoplasm and obvious nucleolus. During the surgery, firstly the lesion was subcutaneously observed on the superficial parotid gland. The mass was oyster white, rubberyand harder than parotid gland. The mass had a capsular appearance differently from the surrounding tissue. Its malignancy was researched by freezing the mass. As a result of pathology, it was in the favor of benign mesenchymal tumor, and the mass was excised with the superficial

parotidectomy. Histologically, the tumor was characterized by monomorphous spindle-shaped cells arranged in a storiform pattern on a background of fibrous stroma and infiltration of surrounding subcutaneous fat and normal salivary gland (Fig. 1). There was little nuclear pleomorphism and low mitotic activity. On immunohistochemical studies, the tumor was stained strongly with CD34 (Fig 2), but negative with SMA. As a result of the pathology, mass was reported as dermatofibrosarcoma protuberans. The patient was followed during 18 months and any relapse was not encountered. The patient was informed about the fact that the follow-up process would take a long time because of the slow course of the disease.

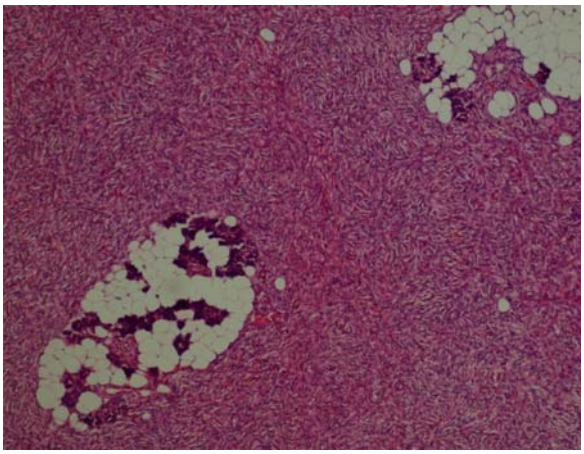

Fig. 1. The cellular spindle cell tumor diffusely infiltrating subcutaneous fat, serous acini and ducts in a storiform architectural pattern (H&EX100).

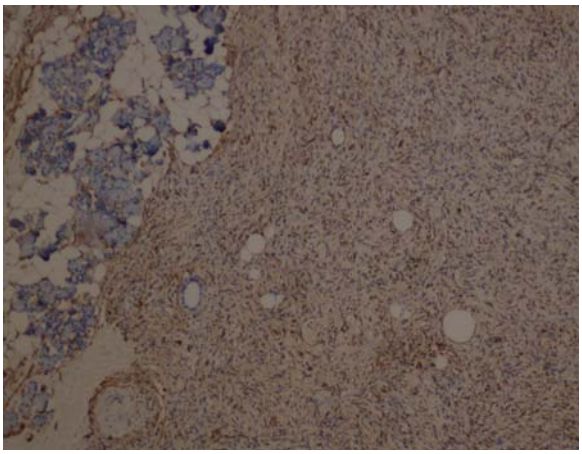

Fig. 2. Diffuse staining with CD34 in tumor cells (Immune-peroxidase X100).

3. Discussion

Dermatofibrosarcoma protuberans was firstly defined to be “progressive and recurring dermatofibroma” by Darier and Ferrand in 1924 (6). Although it is thought that the cellular origin of DFSP is the stem cells in dermis or undifferentiated mesenchymal cells that have neurological, muscular and fibroblastic characteristics (7), the latest immunohistochemical evidences show that they can originate from the dendritic cells (5). DFSP is a rare sarcoma of the skin and its general incidence is 0.8-5/1.000.000 (3). It accounts for less than 0.1% of all malignancies, 2-6% of all soft tissue sarcomas and only 1% of the soft tissue sarcomas of the head-neck region (8,9). Although DFSP can develop in any part of the body, it is most frequently seen on torso (50-60%), extremities (20-30%) and head-neck region (10-15%), respectively (10). Moreover, vulva and parotid gland were reported as the rare localizations for DFSP in the literature (4,6). In our case, the lesion was subcutaneously localized on superficial parotid gland. There was no finding of the lesion on the skin.

In the pathogenesis of DFSP, the fusion between “collagen type I alpha 1 gene (chromosome 17)” generated as a result of (17; 22) (q22:13) the reciprocal translocation and “the growth factor beta chain gene (chromosome 22) secreted from the platelet” is blamed (1). It was thought that this fusion between genes, distortion of the expression of the growth factor beta chain secreted from platelet and the constant activation of the receptor beta protein tyrosine kinase play a role in the development of DFSP (10,11). In the three dimensional reconstruction microscopy is addition to clinical image of DFSP, it is seen that the tumor advances irregularly into the deeps in villous or finger shape. This indicates that the lesion can spread to dermis, subcutaneous and muscle tissue by coursing slowly according to the regions. It is quite difficult to get a surgical space; for this reason, the relapse is one of the most frequently encountered problems (10).

The diagnosis is made by histological and immuno-histochemical assessment. The sampling in pathology is conducted by FNAP or incisional biopsy. While FNAB is enough for the patients whose tumor is thought to recur, an incisional biopsy should be preferred for the appropriate sampling if the patient makes a biopsy performed for the first time (12). In our case, FNAB could not be a guide way, either. Atypical fibroblasts that have histologically hyperchromatic nucleus and eosinophilic cytoplasm are seen as bunches in fusiform. A purl-like appearance created as a

result of the diffuse proliferation of the thick fusiform cells in dermis, hypodermis and intercrossing draws attention. These cells can infiltrate into subcutaneous tissue and consequently causes a honeycomb appearance (2).

The lesion does not have a real capsule; it reaches a fake capsular appearance as a result of the press to the surrounding tissue (2). In our case, the mass made a subcutaneous press and was in a form that can be easily separated. In the separative diagnosis of DFSP, solitary fibrosis, benign fibrous histiocytoma, schwannom, myoepithelioma and other fusiform cell sarcomas should be considered (1,2,6,8,10). Immuno-histochemical analyses are instrumental methods for verifying DFSP diagnosis. Especially CD34 staining is commonly used and its sensitivity was reported to be 84% and 100% (13). While Bcl-2, CD99 and Vimentin are plus positive; CD56, S100 protein, HMB45 and SMA are negative (14). The diagnosis was verified with CD 34 positive, Actin positive and S100 protein negative and SMA negative staining in the immuno-histochemical verification in our case.

The fundamental approach in the treatment of DFSP was surgical. Since the rate of relapse is high after the treatment, an excision or Mohs' micrographic surgery by leaving a surgical space up to 5 cm from the tumor line is suggested (9). In such situations where the broad excision cannot be performed and tumors are close to the surgical space, adjuvant radiotherapy can be given to decrease the risk of recurrence (11).

Although there is a broad excision spaces ranging from 1 to 3 cm in these patients, the rate of recurrence of the lesions in head and neck can be 50% -75% (8,14,15). For this reason, tumor should be excised with its surrounding normal tissue up to 5 cm, subcutaneous fatty tissue and fascia (14). In our case, the mass was excised by the superficial parotidectomy after the pre-diagnosis of the benign mesenchymal tumor. The patients were followed during 18 months and any relapse was not encountered. Mc Peak and his friends (14) encountered the local relapse by 80% after the simple excision in their study. Moreover, Stojadinovic and his friends (15) reported that a surgical space was verified in 80% of the patients with the frozen analysis, whereas positiveness of the surgical margines were too low to accept in 57% of the patients as a result of the wrong negativeness.

They use Mohs' microscopic surgery as the best approach in the treatment of DFSP since DFSP lesions extend in the finger-like protrusion (16). In the Mohs' microscopic surgery applications, the margines of tumor are evaluated three-dimensionally with the sections taken from the frozen or paraffin sections. The tumor is resected with an approximately 2 mm security space and a horizontal section is received from the lower side of the resected mass. The horizontal section is resected until getting a negative margine in the frozen analysis (16). So that both relapse and surgical morbidity can be decreased. When the results were compared, the rate of five years relapse was 13% in the broad surgical resection and any relapse was not encountered in Mohs' microscopic surgery (16). Radiotherapy was only used in some small serials in the treatment of DFSP (17,18). It was reported that positive surgical margins increased 10 years local control of the adjuvant radiotherapy and survival rates in the invasion and recurrence cases (17).

The prognosis of DFSP is generally good. The 5 years survival rate of the patients after the appropriate treatment is between 93% and 100% (15). The metastases are performed with hematogenes and lymphatics and they are generally seen in the late clinical cases or cases where some local recurrences happened. Although DFSP are frequently recurring, their remote metastasis rate is only 1-4% (10). Rutgers and his friends reported 1% regional metastasis and 4% remote metastasis in their DFSP study with 913 patients (8). Mc Peak and his friends detected 4-6% remote organ metastasis (14).

In conclusion, DFSP is quite rare that it was subcutaneously localized with atypically localization on the superficial parotid gland. Moreover, it should be kept in mind that probability of the microscopic spread is high in these patients and they should be followed by reminding that the disease can recur in the following long years.

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