Atypical Fibroxanthoma in Head and Neck

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INTRODUCTION

Atypical fibroxanthoma (AFX) is a malignancy developed typically in the dermis of the head and neck of senior Caucasians, however it is a very rare tumor in Asians [1-5]. In addition, the incidence of AFX in Koreans is very low and only 6 cases developed in the sun-exposed face and hand area have been reported in dermatology [6-10].

Although the metastatic potential is known to be low, it grows rapidly and has a highly local recurrent ability [11]. It must be differentiated from many diseases since it shows the sarcoma pattern consisting of atypical spindle cells within the dermis histology. It can also be considered a benign lesion because there is no pain or local invasion clinically. Although rare, metastases have been reported, and they could not be differentiated from malignant fibrous histiocytoma (MFH) histopathologically. As a treatment, complete excision is best. However, local recurrence can occur frequently because the margin is not distinct when performing an excision. Therefore, a wide excision has been known to be the most effective [4,5]. In this study, we examine a case diagnosed as atypical fibroxanthoma in the head and neck area, and report this case with a review of the literature.

CASE REPORT

Our patient was a 70-year-old male who 3 months ago underwent an incision biopsy at another hospital for a 1.0×1.0 cm sized palpable and protruding reddish nodule in the left infra-auricular area. By result of a histological test, a malignant tumor was suspected and he was transferred to our hospital. In the physical examination, there was a round scar remaining from the previously performed biopsy and the size of the resected tissue was approximately 0.7×0.7 cm. Any other special findings were not detected. The specimen acquired at the other hospital was examined again at our hospital. In our histopathologic finding, we detected a numerously mitotic figure with atypical, bizarre, large and hyperchromatic nuclei (Fig. 1). After immunostaining, we had CD34, CD45, cytokeratin and desmin showing negative reaction with a positive reaction for Vimentin as well as CD68. Finally, we diagnosed it as an atypical fibroxanthoma [2,3]. In addition, the malignancy was confirmed in the previous incision margin, and there was no special findings in a metastatic work-up. Therefore, a wide excision was performed with 2 cm safety resection margin around the lesion which underwent the initial incision biopsy, and there was no malignancy detected by frozen biopsy (Fig. 2A). The defect was then closed by simple suture (Fig. 2B). Twelve months have passed after surgery and the patient is under the follow-up observation at our outpatient clinic without any local recurrence.

Keywords. Atypical fibroxanthoma, Biopsy, Excision
DISCUSSION

An AFX is a rapid growing neoplasm with low metastatic potential, but local recurrence is estimated at between 2%-20% and metastasis has been reported [11,12]. It is a solitary nodule smaller than 2 cm in diameter in most cases and it occurs on the face as well as the back of the hand in Caucasians older than 50 years (average, 70-year-old) in the sun-exposed area [1-5]. It is thought that the length of sun exposure, trauma and previous cutaneous neoplasm history as well as previous radiation history are all involved in the risk factors in the development of AFX. However, the precise cause have not been elucidated [11,13].

Histopathologically, it commonly accompanies ulceration in the epidermis covering the lesion and there is a partial protrusion shape compared with adjacent tissue. Pleomorphic tumors with an indistinct border adjacent to the dermis-epidermis junction are shown. Tumor cells are primarily atypical spindle cells, multinucleated giant cells with a strange shape that cannot be characterized and are also mixed with pleomorphic cells [2]. Many mitosis are observed and cells similar to fibroxanthoma are densely distributed and surround the skin adnexa. With immunohistochemical staining, it is positive for vimentin. However, it shows negative for CD34, cytokeratin, desmin, and S-100 protein [1-5].

Diseases to be differentiated are dermatofibrosarcoma protuberance (DFSP), MFH, leiomyosarcoma, spindle cell cancer, squamous cell carcinoma, etc [3,4]. From the aspect of clinical findings, DFSP is similar to AFX. However DFSP occurs preferentially in the upper limbs and the body of young adults. In addition, DFSP shows the positive for vimentin and CD34. These characteristics would differentiate DFSP from AFX. From the aspect of histological findings, MFH is similar to AFX, however MFH occurs preferentially in the limbs and deep invasion is frequent which is different from AFX. There is also a positive result for Vimentin, and a negative result for CD34 and S-100 protein. Leiomyosarcoma or squamous cell tumors could be differentiated by histologic findings as well as immunohistochemistry test. Recently, the expression of CD99 and S-100A6 has been reported to be a useful immunohistochemical test indicator for the diagnosis of AFX [2,14].

Regarding AFX treatments, although the tumor shows histological malignancy findings, the site of the lesion is the superficial layer in most cases, and the tumor shows a benign course since its metastatic potential is low. It could be treated by complete resection with 2 cm safety resection margin creating a good prognosis. However, the possibility of recurrence is high in incomplete resection cases [11,12,14]. Recurring cases are treated by re-resection or radiation therapy. In general, metastasis is rare. However, in the case of a large tumor or a tumor lacking a distinctive border, the recurrence and metastasis are more frequent [4,15,16].

Our patient had a mass developed in the infra-auricular area which looked like a benign nodule and underwent an incision biopsy. Since it was diagnosed as an atypical fibroxanthoma in

Fig. 1. Histopathologic examination demonstrated pronounced atypia, with bizarre, large, hyperchromatic nuclei numerous mitotic figure (H&E, ×200).

Fig. 2. Intraoperative photograph shows skin excised state about 3×2 cm-sized included previous incisional scar. There are small portion of parotid gland was seen on the base of resection margin (arrow) (A) Simple sutured after wide excision (B).
the histopathologic test, a complete resection was done with 2
cm safe margin around the initial lesion. The postsurgical follow-
up observation period was just 1 year, which may be short for
malignant tumor case. AFX cases would look like a local recur-
rence within 1 year mostly [11]. Therefore we think we can check
the recurrence status every three months for a 2 year period. In
my opinion, the point to be considered in this case is a simple
nodule diagnostic workup. Typically, in this case, a simple inci-
sion biopsy is preferred because of the simplicity of surgery and
less discomfort to the patients. However, we believe it needs to
be an excisional biopsy even if it is a simple nodule, because it
may give rise to more serious problem and we cannot complete-
ly exclude the possibility of malignant tumor.

The authors experienced a case of AFX developed in the head
and neck area. We think this case is very important because, it is
not only a rare case in a Korean, but also consider the question
of the diagnosis of a neck mass once more. As a result, we report
this case together with a review of the literature.

CONFLICT OF INTEREST

No potential conflict of interest relevant to this article was re-
ported.

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