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협부에 발생한 점액양 지방육종의 치험례

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A Case of Myxoid Liposarcoma of the Cheek

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Purpose: Liposarcoma is the most common soft tissue sarcoma, and usually occurs on the thigh or in the retroperitoneal space, but rarely in the oral region. This report presents a case of liposarcoma of the cheek and includes a review of the literatures.

Methods: A 21-year-old woman was admitted with a palpable mass in her cheek of about two years duration, which increased in size gradually initially, but had increased rapidly over the three months. There was no particular pain or tenderness. MRI showed a well-enhanced, well-defined mass, which suspected to be hemangioma.

Results: The spherical, well-encapsulated mass was surgically excised. Biopsy results revealed myxoid liposarcoma. FDG PET-CT on the seventh postoperative day, revealed a minimal to mild FDG-uptake soft tissue lesion around the mass defect area without evidence of distant metastasis. The patient is being observed and undergoing radiation therapy.

Conclusion: Liposarcoma in the head and neck region is a rare disease, and can be overlooked as a benign tumor without a pathologic diagnosis. Therefore, proper treatment and follow-up are required based on an understanding of this disease.

Key Words: Myxoid liposarcoma, Cheek

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I. INTRODUCTION

Liposarcoma is a malignant mesenchymal tumor of adipose tissue and is one of the most common soft tissue sarcomas. Its incidence peaks between 40 and 60 years of age and it is most commonly found in the lower extremities and retroperitoneum. Approximately four percent of liposarcomas are found in head and neck region, and rarely occurs on the cheek. Only three cases of liposarcoma have been previously reported in Korea.

The pathologic classification of liposarcomas is critical for optimal treatment and determining prognosis, and radiation therapy after surgery has been reported to be effective. The authors report the case of a 21-year-old woman diagnosed with liposarcoma by resection biopsy.

II. CASE

A 21-year-old woman was admitted with a palpable mass on the right cheek. The mass became palpable two years previously and had started to grow rapidly during the three months prior to presentation. The mass was round and hard and was not painful during palpation. There was no accompanying any other symptoms and no palpable lymph nodes in the neck.

An MRI performed before admission, revealed a well-enhanced mass with a well-defined margin, which raised suspicion of a benign tumor, such as, a hemangioma (Fig. 1). Surgical resection was carried out under general anesthesia through an intraoral incision in buccal mucosa. The mass was removed without a resection margin. The mass was well capsulated and show no adhesions or signs of inflammation (Fig. 2). Gross observation showed that it was fresh pink, of overall size 6 × 2.8 × 2.5 cm, and was composed of two pieces. It was also surrounded by a soft, thin, fibrous capsule (Fig. 3). The hematoxylin-eosin stain revealed a myxoid stroma lined by fusiform or round cells and a well-developed plexiform capillary network. It also showed lipoblasts in different stages accompanying signet ring cell shaped lipoblasts (Fig. 4).

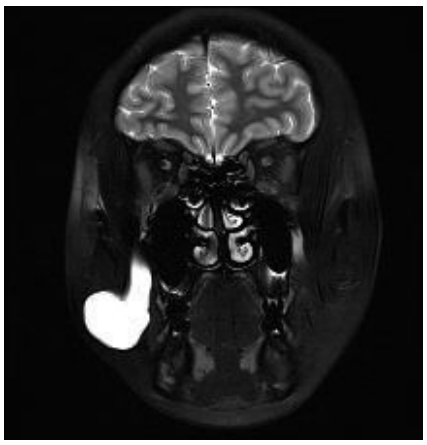

Fig. 1. Preoperative MRI image finding. Enhanced ovoid, well demarcated mass is seen on the cheek.

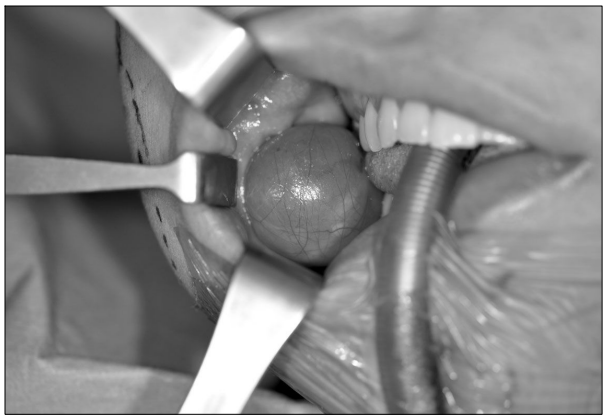

Fig. 2. Intraoperative finding of myxoid liposarcoma of the cheek. Smooth, vacularized and capsulated mass is seen through an intraoral incision in buccal mucosa.

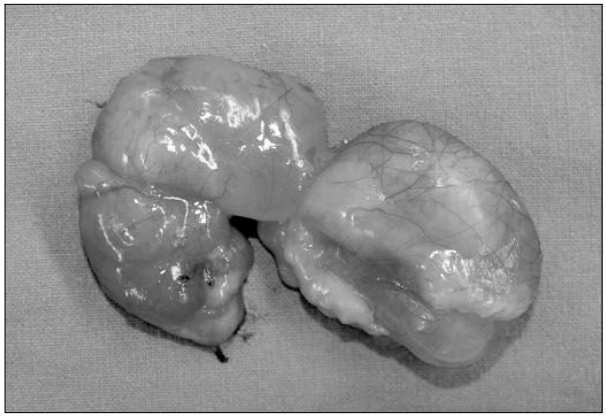

Fig. 3. Gross findings of specimen. Ovoid & capsulated mass is seen.

And the tumor cells showed a strong immune reaction to S-100 proteins (Fig. 5). The patient was diagnosed with

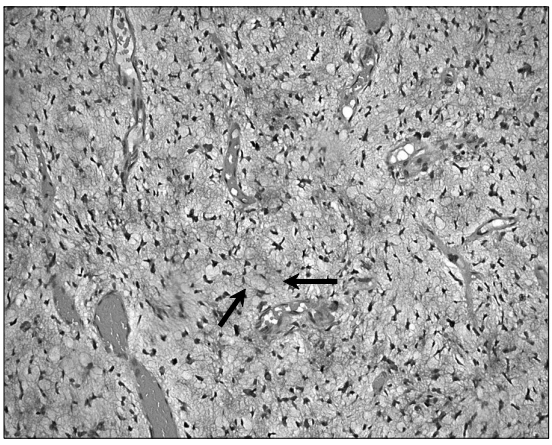

Fig. 4. At some focus, plexiform capillary network, large number of lipoblasts at varying stages & signet ring cell shaped lipoblasts (arrows) were present (Hematoxylin and eosin stain, × 200).

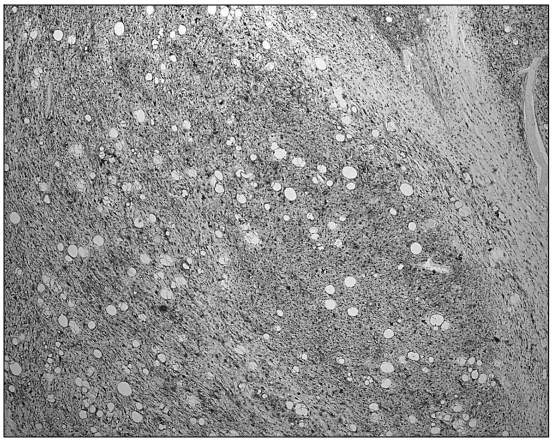

Fig. 5. Microscopic feature of myxoid liposarcoma. The tumor cells showed strong immunoreactivity for S100 protein.

myxoid liposarcoma. No complications accompanied surgery and the patient was discharged at three days postoperatively. An FDG PET-CT taken at 7 days postoperatively showed mild uptake in previous operation site, but no signs of distant metastasis. The patient underwent 32 radiotherapies (6000 cGy) and showed no signs of metastasis on FDG PET-CT four months later, exhibiting signs of complete remission of the previous lesion.

III. DISCUSSION

Liposarcoma is one of the most common malignant mesenchymal tumors with an onset peak in 40 to 60 year old. Cases of young adults and teenagers have been reported, but cases of liposarcoma in children are rare.[1-5]

Genetics factors, trauma, infection, and exposure to radiation have been implicated to be causative factors, but no well-established factor has been identified.[3,4]

Liposarcoma occurs about 1.5 times more frequently in men but shows no associations with race or geography. The retroperitoneal area and lower extremities are common sites of origin.[1,3,4,8]

Liposarcomas account for 15% of all soft tissue sarcomas, but only 4% of liposarcoma occur in the head and neck region.[8] Enzinger and Weiss reported 60 cases of liposarcomas in the head and neck region among 1,067 cases, Noriaki reported treating 18 cases of head and neck liposarcoma from 1970 to 1996, and Eleni et al. reported 45 cases of intraoral liposarcoma from 1944 to 1999.[1,2,6] Only four cases, including the present case of head and neck liposarcomas have been reported in Korea. In the head and neck region, location have reported to be; cheek (37%), maxilla (26%), palate (16%), mouth floor (11%), and mandible and tongue (5%).[1]

Symptoms depend on tumor size and location, and no symptoms are apparent until the mass is large enough to pressurize adjacent tissues. Therefore, diagnosis is often delayed and the presenting symptoms may include tenderness, pain, and functional disturbance.[2,3,5]

Liposarcomas are usually well-circumscribed and encapsulated and the cut surface may be mucinous, gelatinous, or firm, depending on the pathologic composition of the tumor. In particular, the myxoid type of liposarcoma shows various stages of lipoblast differentiation, a plexiform capillary network, and diverse myxoid stroma, and strong immunoreactivity to S-100 protein.

The classification of liposarcomas is diverse and depends of the amount of differentiation and the cell type. The most commonly used classification was proposed by Enzinger and Weiss, which was also used to classify liposarcoma in the described case. This classification describes five types: well-differentiated, myxoid, round cell, dedifferentiated, and pleomorphic. Myxoid liposarcoma is the most common form.[1,4-6]

Prognosis depends on pathology, location, and the appropriatness of surgical treatment. And the five year survival rate differs in the descending order, follow the pattern well-differentiated, myxoid, pleomorphic, round cell type.[1,3-7] Less differentiated, more polymorphic tumors, and tumors of deeper origin have been shown to have higher frequencies of recurrence and metastasis.[2] Well-differentiated and myxoid types have been reported to show lower frequencies of metastasis, but more prone to local recurrence.[2,5] In recurred cases, the tumor pathology usually matches that of the primary tumor, but cases with a more malignant tumor have been reported.[3] Metastasis is usually hematogenous and commonly occurs in the lung. But metastasis to lymph nodes is rare.[1]

Lipomas do not develop into liposarcomas, and liposarcomas do not usually develop from other benign lesions, although such cases have been reported.[3]

The most effective treatment is surgical excision of the tumor with a generous resection margin. The combination of surgical excision and radiotherapy has been reported to reduce recurrence, but chemotherapy has not shown to be helpful during the treatment of liposarcoma.[1-8]

Liposarcoma in the head and neck region is a rare disease, and can be overlooked as a benign tumor without a pathologic diagnosis. In this case, the authors did not have possibilities of liposarcoma in mind since the lesion had a clear resection margin and did not invade the muscular layer. We emphasize that proper diagnosis, treatment and follow-up are required based on an understanding of this disease. Therefore, we authors present a case of myxoid liposarcoma of the cheek of a 21-year-old woman with a review of literatures.

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