ANGIOLYMPHOID HYPERPLASIA WITH EOSINOPHILIA : A CASE REPORT

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Angiolymphoid hyperplasia with eosinophilia is an unusual and controversial lesion that occurs primarily in the head and neck area. This lesion was usually confused with Kimura's disease. We present the case of a 32-year-old woman with massive soft movable mass in left facial area which was diagnosed preoperatively as a fasciitis nodular. The final histologic diagnosis of the excised mass was angiolymphoid hyperplasia with eosinophilia(ALHE).

Key Words : Kimura's disease, ALHE.

I. INTRODUCTION

Angiolymphoid hyperplasia with eosinophilia (ALHE) is a clinicopathologic entity that shows spectral variation in its clinical and microscopic presentations[1]. Histologically, ALHE[2] is characterized by two components : a proliferation of hyperplastic vessels lined with plump endothelial cells and an inflammatory infiltrate comprising lymphocytes and eosinophils[1].

In 1948, Kimura and associates described a benign disease in young men characterized by nodular subcutaneous infiltrates of lymphocytes and eosinophils, often with associated regional lymphadenopathy and generally with blood eosinophilia[2]. ALHE is currently regarded as a different entity altogether, a reactive and not a neoplastic process, related to autoimmune disorders with unknown pathogenesis[3,4].

The aim of the present study is to give a review of the literature and to discuss differences between Kimura's disease and ALHE.

II. CASE REPORT

A 32-year-old woman was admitted to the department of oral and maxillofacial surgery, Chosun University Dental Hospital in April, 1993, because of massive painless swelling of left supraorbital, preauricular, buccal, and submandibular area. The patient noticed the swelling 1 year earlier, and then recurrent swelling was repeated periodically, but she ignored it. There was no past history of any trauma or infection. She has been suffered from chronic renal insufficiency. Family history was unremarkable. Clinical examination revealed a well-circumscribed enlargement in left supraorbital and parotidomasseteric region, and the soft, movable

mass measured 10×10cm in parotidomasseteric area and 2×2 cm in supraorbital area(Fig. 1). Neither excretory disturbance of saliva in the parotid gland nor evidence of xerostomia was obvious. The sialogram of left parotid gland revealed normal finding(Fig. 2). In aspiration, no fluid was withdrawn. Infectious source was never detected. There was no cervical lymphadenopathy. Generalized dark pigmentation was seen on overlying skin of body and extremities. Her admission workup disclosed 14, 760 counts of WBC. There was blood eosinophilia (62.4%). Tentatively we diagnosed neurofibromatosis, parotid gland tumor, or infectious lesion of unknown origin. We performed incisional biopsy. Pathologic diagnosis was fasciitis nodular. We planned completely surgical excision and excisional biopsy.

In CT finding, Iso or slightly low density homogenous huge well-circumscribed mass was seen laterally to the masseter muscle, parotid gland and sternocleidomastoid muscle. Extension of mass to submandibular area inferiorly and to the periorbital area superiorly were seen. There was no demonstrable definitive abnormal bony destruction or erosion(Fig. 3).

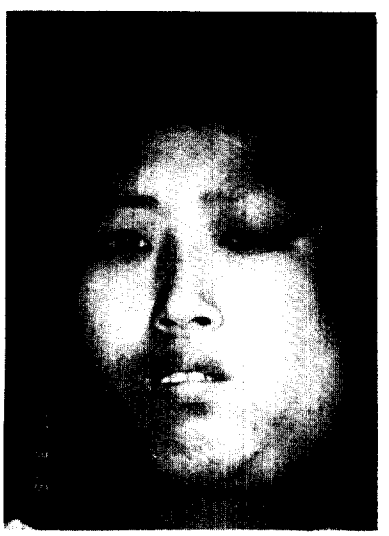

Fig. 1. Preoperative photograph. Well-circumscribed soft, movable mass in left supraorbital and parotidomasseteric area.

Fig. 2. The sialogram of left parotid gland reveals normal finding.

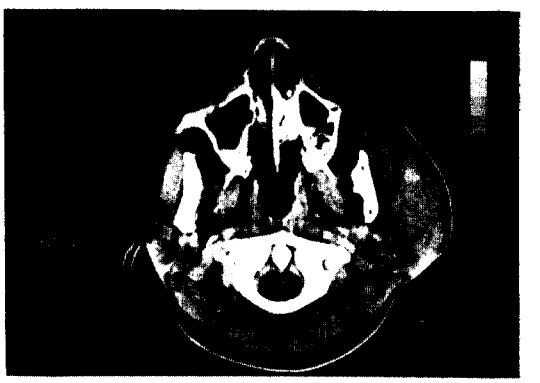

Fig. 3. Horizontal CT finding. Homogenous huge well-circumscribed mass is seen laterally to the masseter muscle and parotid gland.

She was admitted in internal medicine for about 3 weeks in order to treat the CRI. And then she was transferred to our department for surgical excision of mass.

At operation, the mass which was irregularily outlined from the surrounding tissues was excised and superficial parotidectomy was carried out because of indurated gland architecture(Fig. 4).

Histopathologically, clumps of lymphocytes and

Fig. 4. Operative finding. Irregularily outlined huge mass is being excised. Lymph node enlargement is seen.

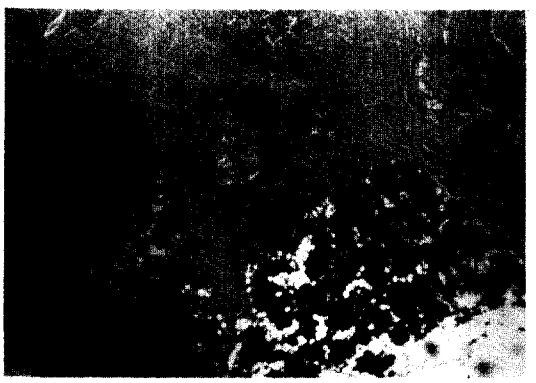

Fig. 5. Photomicrograph showing dilated intercalated ducts partly replaced by sheets of chronic inflammatory cells(Arrow). (H-E, ×100)

eosinophils interspersed by fine delicate fibrous tissue were seen. In serous acini, dilated intercalated ducts partly replaced by sheets or clumps of chronic inflammatory cells infiltration. In several lymph nodes, swollen or hyperplastic lymphoid tissues with plenty of eosinophilic infiltration were identified (Fig. 5, 6). Final pathologic diagnosis was angiolymphoid hyperplasia with eosinophilia.

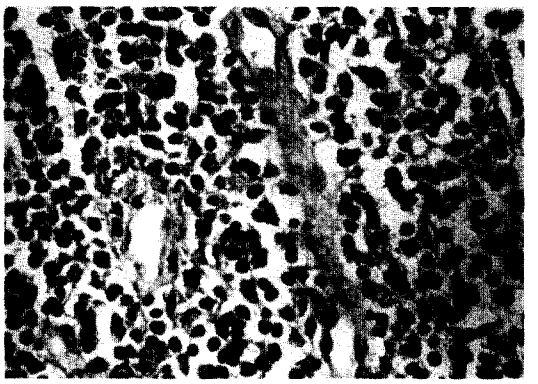

Fig. 6. Clumps of lymphocytes and eosinophils interspersed by fine delicate fibrous tissue is seen. (H-E, ×400).

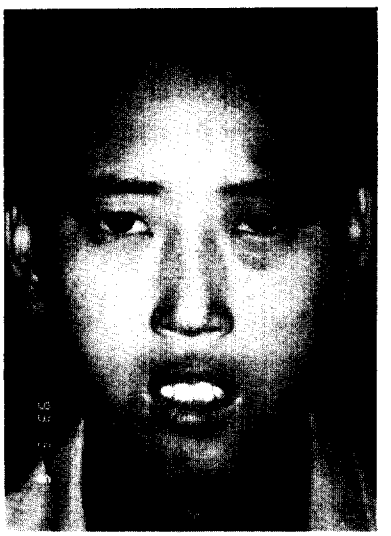

Fig. 7. Postoperative photograph.

She has been followed-up for 3 months and has a favorable prognosis. She will have a long-term follow-up because of a possibility of recurrence(Fig. 7).

III. DISCUSSION

Angiolymphoid hyperplasia with eosinophilia is a rare, benign angioproliferative lesion with a predilection for the facial skin[5]. ALHE is an unusual and controversial lesion that occurs primarily in

the head and neck area ; oral involvement is rare[2].

Some authors suggested that ALHE is equivalent to Kimura's disease[6,7,8,9]. Rosai[10] considered ALHE a separate entity from Kimura's disease which shows features more suggestive of a reactive process. ALHE is a vascular neoplasm with proliferation of a atypical cells. Peters et al[2] suggested that different entities, which include vascular neoplasms, Kimura's disease, and possibly other reactive conditions, may be encompassed in the Western literature as ALHE. Angiolymphoid hyperplasia with eosinophilia is the most frequently used term for the disease in non-oriental persons. Thomas et al.[11] suggested that we prefer to retain the entity of Kimura's disease as part of the spectrum of ALHE until more definitive evidence becomes available.

Iguchi et al.[12] studied Kimura's disease and its relation to angiolymphoid hyperplasia with eosinophilia. Kimura's disease, eosinophilic granuloma of soft tissues, is chronic inflammatory condition and appears as subcutaneous tumor-like nodules in the head and neck regions. Histopathologically, it is characterized by the presence of lymphoid follicles, marked infiltration of eosinophils, fibrosis and vascular proliferation. And it has male predominance and geographical peculiarity. This lesion usually developes in deep regions of soft tissue and tend to be larger than those of ALHE. It has higher values of eosinophilia : 30~40% is common, sometimes 80%. But ALHE has female predominance. ALHE usually developes in superficial regions of head and neck. Histopathologically, it is characterized by a few lymphoid follicles, remarkable proliferation of endothelial cells, mast cells and no fibrosis. We consider Kimura's disease and ALHE to the same entity. It is considered that our case belongs to late stage of ALHE.

Similar lesions were also known as "inflammatory angiomatous nodule"[13], "pseudo or atypical pyogenic granuloma"[14], papular angioplasia"[15], "subcutaneous angioblastic lymphoid hyperplasia with eosinophilia"[6], "histocytoid hemangioma"[16,17] and "nodular angioblastic hyperplasia with eosinophilia and lymph folliculosis"[18].

Past reports emphasized pruritus as the most common symptom associated with ALHE and pain as an uncommon symptom[19]. In contrast, Thomas et al.[11] found pruritus in only 15% of patients reporting symptoms and pain and lesional growth as the most commonly reported symptoms, in 31% and 39% of patients, respectively. Spontaneous bleeding and pulsation over certain lesions of ALHE were consistent with the vascular nature of the lesion. Peripheral blood eosinophilia and lymphadenopathy associated with ALHE were reported too sporadically. Our patient's sign and symptoms were periodic recurrent painless swelling and peripheral blood eosinophilia. Peters and his colleagues reported the analyses of total 102 cases of ALHE. It usually developes in third to fifth decades. It has female predominance. The lesions were restricted to the head and neck area in 86% of the cases. The smallest lesion measured 0.2cm, and the largest measured 6cm. Regional lymphadenopathy was noted in 11% of the cases. 44% of the cases was multiple lesion. Blood counts were reported in 53 cases ; there was an eosinophilia greater than 4% in 39% of the cases and greater than 10% in 23% of the cases[2]. In our case, there was an eosinophilia of 62.4%.

For the most part, the pathogenesis is unknown, with opinion divided as whether the lesion represents a true vascular neoplasm or a reactive healing phenomenon following a variety of insults such as trauma, infection, or hormonal imbalance[11]. Many hypotheses include infection, allergy, trauma, hormonal imbalance, an overgrowth of an atypical population of endothelial cells, and inflammatory skin disorder[11,20,21,22]. Lavinia et al.[23] reported that the lesions developed shortly after immunization. The role of renin-angiotensin cascade in simulating vessel proliferation was also proposed by Fernandez et al[24]. The presence of either AV shunt formation

or intravascular endothelial proliferations in lesions associated with trauma and pregnancy suggests a possible relationship of trauma and pregnancy to the pathogenesis of ALHE[2,11].

Histopathologically, two characteristic features are vascular and inflammatory components. The vascular component tends to dominate in early or active lesions, whereas older or quiescent lesions display the more chronic inflammatory or lymphoid characteristics. Vascular component consists of small vessels with prominent and pleomorphic endothelial cells, and the cellular component consists of an inflammatory infiltrate comprising lymphocytes, eosinophils, and histiocytes[2,25]. Wells and his collegues[19] emphasized that the essential histologic components of the lesion are : (1) vascular proliferation with an angiomatoid proliferation of capillary vessels, and solid masses of endothelial cells appeared in several cases ; (2) marked tissue eosinophilia at all stages ; (3) numerous mast cells ; (4) diffuse lymphocytic foci ; (5) lymphoid follicles appeared in many cases ; and (6) no fibrosis. Regional lymph node involvement biopsy reveals reactive hyperplasia. In our case, Normal saliva excretion and normal sialogram finding were seen but parotid gland infiltration was confirmed in operative and pathologic finding. And active vascular proliferation with plump endothelial cells was rarely identified. There was no regional lymphadenopathy.

The differential diagnosis is extensive and includes pyogenic granuloma, granuloma faciale, lymphocytoma, persistent reaction to insect bite, angiosarcoma, eosinophilic granuloma, angiomatous lymphoid hamartoma, arteriovenous malformation, hemangioma, lymphangioma, nevus, and folliculitis. The most significant lesion to be differentiated is angiosarcoma because of the need for extensive resection. In ALHE, no anaplasia was present and the mitotic figures were noted with a frequency of 1 per 20 high-power fields[1,18,25,26]. The authors suspected neurofibromatosis and fasciitis nodula. Fasciitis nodular(pseudosarcomatous fibromatosis) is a reactive fibroproliferative lesion extending as a solitary nodule from the superficial fascia into the subcutaneous fat or less frequently, into the subjacent muscles. Incisional biopsy specimens showed diffusly infiltrated chronic inflammatory cells through out the field and in part some lymph follicles with hyperplastic changes were noted. Neurofibromatosis was suspected because of generalized dark skin pigmentation and soft massive movable mass.

The treatment used and recommended by most authors is complete surgical excision. Deep surgical excision had the most favorable result with no recurrences[2,26,27]. Others have used carbon dioxide and argon lasers, intralesional or systemic corticosteroid, radiotherapy, cytotoxic chemotherapy, curettage and electrodesiccation, and cryotherapy[25,26,28]. Peters et al. suggested that treatment of choice is surgical excision but excision in combination with electrocautery, radiotherapy, or corticosteroid therapy is necessary in multiple recurrent lesions[2]. Olsen et al.[11] reported spontaneous resolution.

Recurrences are not uncommon ; they have been reported in up to 33% of patients, but are most frequent when blood eosinophilia is present[1,2,12]. Recurrence of ALHE is a clinical problem, with up to one third of patients having a recurrence in the same anatomic site months to years after initial therapy, even when surgical margins were said to be free of lesion after excision[29]. In Peters and his colleague's review of all cases without eosinophilia, recurrences were not found in spite of the fact that the study included patients with multiple lesions[2]. No metastasis was found[11]. In Korean Oral and maxillofacial surgical field, 3 Cases with ALHE were reported. Three cases developed at facial area[8,9,30]. Our patient has a possibility of recurrence because of blood eosinophilia. Long-term follow-up is necessary. If there were recurrence, we would perform a adjunvant therapy ; such as surgical excision in combination with radiation or corticosteroid therapy.

Ⅳ. CONCLUSION

We had following conclusions with peculiar case report and literature review.

1. The diagnosis of ALHE prior to biopsy is difficult because of its nonspecific morphologic features.
2. We consider ALHE a same entity as Kimura's disease.
3. Treatment of choice of ALHE is surgical excision. But recurrence is always possible. If there were recurrence, combination therapy should be considered.

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국문초록

호산구 증가증을 동반한 ANGIOLYMPHOID HYPERPLASIA의 증례보고

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호산구 증가증을 동반한 ANGIOLYMPHOID HYPERPLASIA는 주로 두경부의 표층에 발생되고 진단에 어려움이 있는 특이한 질환이다. 일본에서 처음 보고된 이래 주로 동양에서 종종 보고되어온 KIMURA'S DISEASE와 많은 논란이 되고 있다.

저자등은 좌측 안면부에 거대한 유동성의 종괴를 주소로 내원한 환자로서 술전 진단에 상당한 어려움이 있었고 술후 최종적으로 ALHE로 진단된 32세 여자환자를 문헌고찰과 더불어 보고하는 바이다.