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A CASE REPORT OF MELANOTIC NEUROECTODERMAL TUMOR OF INFANCY

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The melanotic neuroectodermal tumor of infancy(MNTI) is a rare childhood neoplasm with a clinical presentation. Because of its rapid growth pattern and bone resorption, the lesion can be mistaken for a malignant neoplasm. Although an aggressive growth rate and radiographic appearance, the MNTI almost always behaves in a benign fashion and can be treated with total excision.

We presented the MNTI occured in the left maxillary alveolar ridge of 5 month old female infant, showing bluish enlargenent of alveolar mucosa with the displacement of central decidious incisor. And after the surgical excision of the mass, there is no recurrent tendency.

I. INTRODUCTION

Melanotic neuroectodermal tumor of infancy (MNTI) is an uncommon, almost invariably benign neural crest origin neoplasm that was first described in 1918 by Krompecher. As the name implies, it is found predominantly in infancy : 82% of patients are 6 months or less of age, and 92% are under 12 months[1,3,6,13,14].

The predominant site of origin is the premaxilla, where nearly 80% of the reported cases have occurred. The remainder arise in other locations, but even with these, there is a predilection for the head : the skull and mandible. Infraclavicular sites of origin include the epididymis, thigh, femur, mediastinum, and shoulders. An axial presentation is nearly always evident. Males and females are equally affected.[1,3,6].

Clinically, intraoral examination revealed a large mass approximately 3×4cm size centered on the premaxillary area, and bony destruction appearance[1,3,6].

Almost cases of radiographic examination revealed a destructive, poorly demarcated slightly radiopaque mass with a pattern reminiscent of the "sunray" appearance associated with osteosarcoma[3,6].

Especially, because high urinary levels of urinary VMA(vanilmandelic acid) are common findings in the individuals with neural cell tumors such as pheochromocytoma, ganglioneuroblastoma, neuroblastoma, and retinoblastoma, MNTI is thought to be neural crest originated tumor[5,14].

Histologically, the tumor is composed of a nonencapsulated mass of dense fibrovascular tissue containing small nests and compressed cords of basophillic tumor cells. Some of the cells have small, round, hyperchromatic nuclei and scanty cytoplasm reminiscent of neuroblasts, while others have larger, round, less chromatic nuclei and cytoplasm stuffed with melanin[5,6,12,13].

The treatment is surgical excision of tumor mass together with removal of adjacent teeth and there is relatively lower recurrence or metastasis rates[1,3,6,7].

So, we present the case report and literatures about the melanotic neuroectodermal tumor of infancy.

II. CASE REPORT

A 5 month old female infant came to the Department of Oral and Maxillofacial Surgery, Kyung Hee University Dental Hospital for the evaluation of a slowly growing left upper lip area from 2 weeks ago.

Clinically bluish tinged submucosal firm mass, which elevated the left upper lip and labial area, was palpated on the upper left alveolar bone, and anterior hard palatal area(Fig. 1).

At first pigmented blood like fluid was aspirated, so there was no I and D, but mucosal biopsy on the swollen alveolar crest area was practiced about 3㎜×3㎜. As a result of immediate frozen biopsy its epithelial cell was looked like malignancy, and after 3 days, the biopsy presented melanotic neuroectodermal tumor of infancy(Fig. 2).

1 week later, rapid tissue growing pattern on the biopsy area was appeared, and accompanied by the increased urinary secretion of 3-methoxy 4-hydromandelic acid(VMA). Under the general anesthesia, the broad surgical excision was done on the alveolar crest area including the surround tissue and hard palate area, but there was no puncture of left maxillary sinus wall(Fig. 3).

After surgical excision, the defect area was packed with nitrofrazone gauze and supported by the suture, and there was no daily dressing. 2 weeks antiobiotics and supportive therapy was practiced, and discharged with the elemination of packing nitrofrozon gauze. We found the mucosal covered defect area after 4 weeks. And then there was no another recurrence evidence(Fig. 4).

Grossly, the submitted specimen was brownish black fibrotic mass, measuring 3㎝×2.5㎝×2㎝ and including 4 deciduous teeth. Cut surfaces of tumor mass revealed on almost entirely black pigmented area in the background of white yellow fibrotic tissue, and gave rubbery consistency(Fig. 5).

Microscopically numerous nests of tightly packed immature cells, surrounded by a proliferative, fibrous stroma containing large, melanin laden cells were found throughout the tumor mass. Nests of tumor were arranged in an alveolar pattern with central cells being exceptionally immature, basophilic, and with little cytoplasm. Moderate mitotic activity was noted within the nest. Peripheral cells appeared to be undergoing a process of differentiation and en-

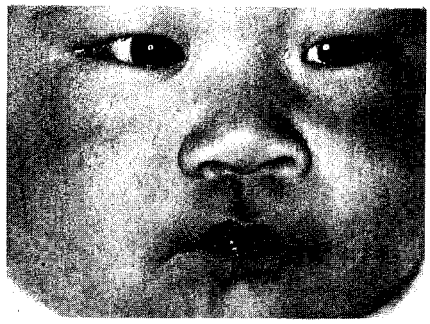

Fig. 1

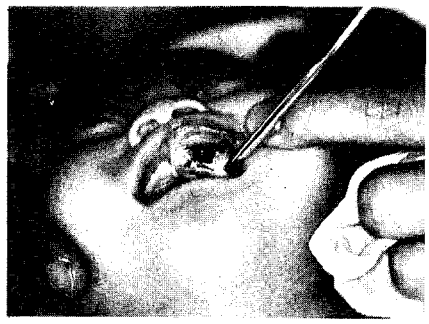

Fig. 2

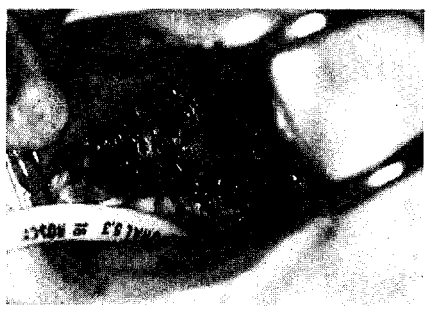

Fig. 3

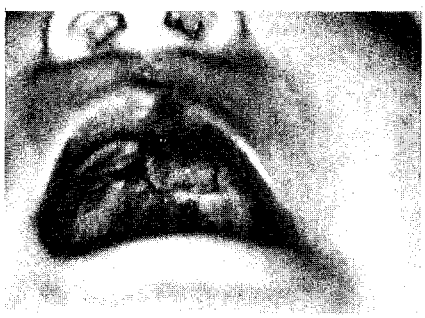

Fig. 4

Fig. 5

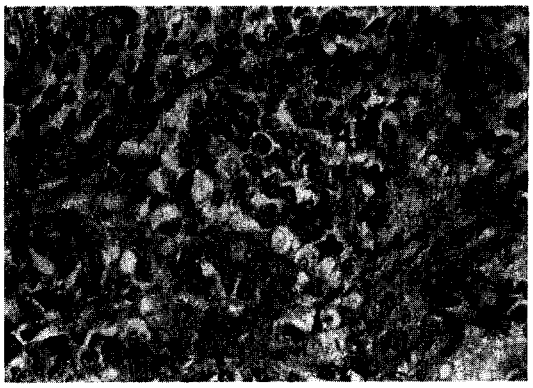

Fig. 6

largement due to increased cytoplasmic volume. The latter cells contained abundant melanin and appeared to proliferate into surrounding stroma assuming a cuboidal shape(Fig. 6).

III. DISCUSSION

MNTI is a benign neoplasm which has stimulated a long debate and controversy as to its histogenesis. The first case to be reported in the literature was disignated a "congenital melanocarcinoma" by Krompecher in 1918. He described a pigmented tumor of the maxilla associated with a developing tooth and elements of dental lamina in a 2-month-old infant[1,3,8].

In 1926, Mummery and Pitts reported a case of pigmented maxillary tumor in a 6-month-old female. The characteristics of the tumor suggested that it arose from some aberration of dental epithelium, and the term "melanotic epithelial odontoma" was induced[8].

In 1947, a tumor reported by Halpert and Patzer contained pigmented epithelium that was suggestive of the ciliary body of the eye. Small unpigmented cells that resembled neuroblasts from retinal neuroepithelium were also present. They suggested that the tumor arose from the entrapment of the retinal anlage in the embryonic fusion lines of the developing maxilla[1,3,6].

In 1957, Shultz reported a malignant melanotic tumor of the uterus, but there is controversy surrounding the histogenesis. Some authors consider this uterine tumor as a malignatnt teratoma with melanin[16].

In 1966, Borello and Gorlin reported a case of melanotic tumor in the maxilla of a 3-month-old boy. Prior to surgical removal of the tumor, there was increased urinary excretion of 3 methoxy 4-hydroxymandelic acid(VMA). After the tumor, was removed, the VMA excretion returned to normal. Since high urinary levels of VMA are common in other tumors of neural crest origin, Borello and Gorlin belived that this was highly suggestive of neural crest origin. They recommended the name "melanotic neuroectodermal tumor of infancy" for this tumor[1,3,5,8,13].

In 1974, Stowens and Lin reviewed the literature and accepted seventy seven examples of this tumor. Their data showed no sexual predilection of the tumor. Although the maxilla was the most common location, 30 percents of the tumors arose in other locations. of this 30 percents, only seven did not occur in the head region[1,3,7,8].

In 1980, Block and Wait represented the first report of metastasis and death from a melanotic neuroectodermal tumor of infancy developing in a previously healthy infant. There was nothing in the clinical presentation or the histologic appearance of the original lesion to indicate the fatal outcome. After the first excision there was no evidence of recurrence for 18 months, until an infraorbital mass appeared and was biopsied. This proved to be a recurrent melanotic neuroectodermal tumor. The second lesion was treated by a partial maxillectomy, which was followed by recurrence and a more rapidly growing tumor. A hemimaxillectomy was performed and was quickly followed by contralateral lymph node metas-

tasis and facial recurrence. The tumor failed to respond not only to three surgical procedure but also to high dosage irradiation and multiple chemotherapeutic agents. At biopsy, the tumor resembled a neuroblastoma in its histologic appearance and distribution. From the above report, it is evident that these tumors may have the potential to behave in malignant fashion[8].

The primary treatment of conservative surgical excision should not be abandoned, however, when more rapidly growing recurrences were occurred, the potential for malignant degeneration should at least be considered.

Ⅳ. SUMMARY

A clinicopathologic and literature review of the melanotic neuroectoderaml tumor of infancy is presented. In the site of predilection, the maxila, the tumor is consistent in its clinical presentation and histopathologic apperance. Although MNTI may be biologically benign, documented examples of malignant cases dictate that MNTI must be approached with circumspection.

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국문초록

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저자등은 유전치의 전위와 함께 상악좌측 치조 점막의 종창을 주소로 내원한 5개월된 여아에서 발생한 유아성 흑색 신경외배엽성 종양을 치험하였으며, 술후 양호한 치료 경과를 보이고 있기에 보고하는 바이다.