HAND — SCHULLER — CHRISTIAN DISEASE

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SUMMARY
Hand-Schuller-Christian disease is a chronic disseminated form of Hystiocytosis-X in which there is minimal or moderate visceral involvement in addition to bone lesions. The manifestations of the disease usually begin at two or three years of age. The commonest site of origin is the intraosseous reticulum and its derivatives. In this paper a case of Hand-Schuller-Christian disease and its clinical and radiological findings have been presented.

Key Words: Hand-Schuller-Christian disease, Hystiocytosis-x.

INTRODUCTION
Known as a congenital disease of the reticuloendothelial system (RES), Hand-Schuller-Christian (HSC) disease begins in childhood and continues thereafter. The typical lesions are seen in the skull, in the membranous bones and to a lesser extent in the long bones (1,2).

It's characteristic signs are defects in the skull, diabetes insipidus and exophtalmos. Splenomegaly, skin pigmentation, endocrine disorders and sometimes hypercholesterolemia may be seen (3,4). It is classified as one of the diseases of the hystiocytosis-x group and its symptoms and signs begin in the 2 nd and 3 rd years of life. The severity of the disease varies from patient to patient (1,2,4).

Although the etiology is still unknown, the disease is thought to be a disorder of the metabolic control of intracellular lipid function. Congenital enzyme defects or infectious agents such as viruses have also been suggested as a cause (1).

Well limited large defects and cortical narrowing may be seen in the skull and in the membranous and long bones. There is no periosteal reaction and the defects are mostly multiple (1). In many cases the maxilla and the mandible are also involved and the teeth may become loose and drop out (5). The lesions are sensitive to x - ray. After radioteraphy ossification can be seen in lytic areas.

Diabetes insipidus can be controlled with extracts of the posterior lobe of the pituitary gland. Developmental and sexual disorders can be treated with anterior lobe extracts. Corticosteroids also have an important role in the pharmacoteraphy of the disease (1).

In this paper a rare case of HSC disease has been presented.

CASE REPORT
The patient (E.K., 3 ♂) was admitted to Alanya State Hospital with the complaint of pain in his fourth molar teeth. Upon physical examination no physical abnormalities were detected other than splenomegaly extending 2 cm. below the costal margin, and an increased degree of irritability.

On radiological examination, the bony structure and bone age correlated with his chronological age. The iliac apophyses were not visualised and the Y cartilage of the acetabulum and the ischial cartilage were open. Wide lytic areas were detected at the right iliac wing (Fig. 1).

Skull x - ray revealed large lytic lesions in both parietal bones, and the same lesions were also seen at the right mandibular corpus (Fig. 2). The 4 th and 5 th upper molar teeth appeared to be hanging in the air (Fig. 3). This finding is pathognomonic of HSC disease.

DISCUSSION
The first lesions of HSC disease are seen at the intraosseous reticulum and its derivatives. If the disease is not severe the lesions are only present in the bones. In severe cases which present earlier and have malignant course, many organs such as the lungs, brain, heart and kidneys may be affected by the disease. Rarely the disease has a latent course with a solitary lesion, but this type may also progress to the typical multicentric form.

The classical triad of skull lesions, diabetes insipidus, and exophtalmos is seen only in 10 % of cases (1, 2). Lesions of the sella and base of the skull with releated pituitary dysfunction are seen in 1/3 of cases. If the posterior lobe of the pituitary gland is affected, the signs of diabetes insipidus may be seen (1, 2).

The case presented, demonsrates the typical osseous lesions of the early period of this disease.

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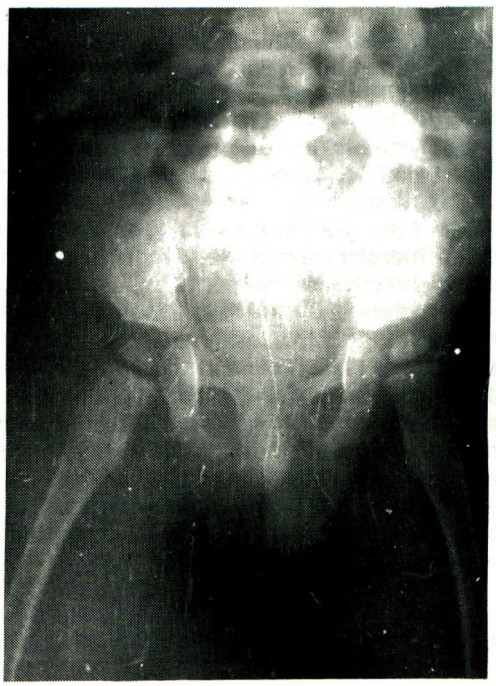

Fig. 1.

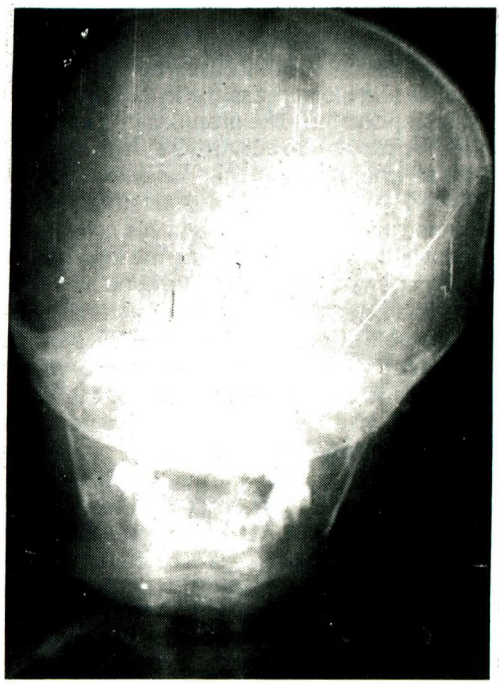

Fig. 2.

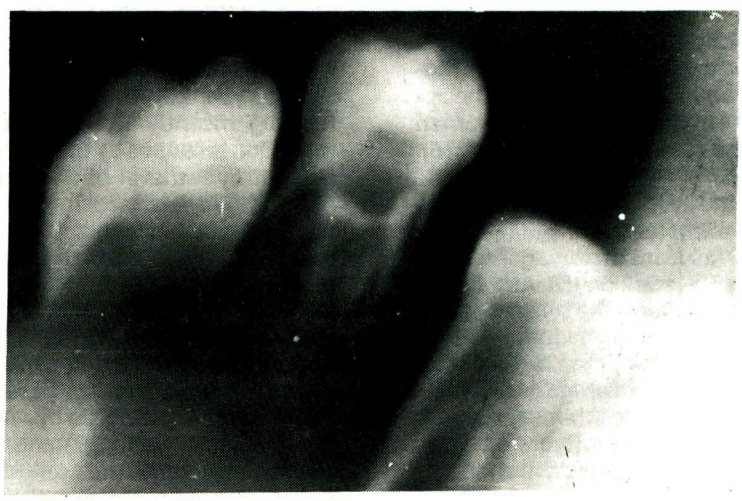

Fig. 3.