Otolaryngological Complications of Osteopetrosis

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Abstract

Osteopetrosis is a rare inherited bone disease that affects both humans and various mammals. The authors report on two cases of osteopetrosis with otolaryngological complications. One patient had the childhood form and presented with chronic otitis media and brain abscess. The second patient had the adult form and presented with sinusitis from tooth extraction which developed into chronic osteomyelitis of the maxillary bone.

Key word : Osteopetrosis, Chronic Otitis Media, Brain Abscess, Osteomyelitis, Maxilla

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Osteopetrosis is a systemic bone disease. It was first recognized in 1904 by Albers-Schonberg (1). The disease is the result of a defect in the ability of the osteoclasts to resorb bone and mineralize cartilage(2). Two forms of the disease occur, a severe form occurring in children and a more benign form affecting mainly adults. The childhood form has an autosomal recessive inheritance and is usually fatal before the second decade of life(2,3). The clinical manifestations of concern to the otolaryngologist include nasal obstruction, deafness, and stenosis of the neural foramina, which can lead to optic atrophy, and oculomotor and facial palsies(4). The adult form is usually autosomal dominant, but it is sometimes

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autosomal recessive. Herein the authors describe the complications of two cases of osteopetrosis: one had the childhood form and the other the adult form.

CASE REPORTS

Case 1

An 18-year-old female was diagnosed as having osteopetrosis at the age of seven years when she developed recurrent osteomyelytis of the mandible, bilateral optic atrophy and complete right facial palsy. Sequestrectomy, curettage and antibiotics remedied the condition. However, at 15, the patient suffered right chronic otitis media which was treated with mastoidectomy in Loei Provincial Hospital, Thailand.

On her last admission to Srinagarind Hospital she had right chronic otitis media with cholesteatoma, right postauricular fistula (Fig. 1) and a brain abscess at the right cerebellar hemisphere (Fig. 2). Treatment included a ventriculoperitoneal shunt, a right radical mastoidectomy and parenteral antibiotics for six weeks. After treatment, the cerebellar abscess had decreased in size and there was no longer any discharge from the right ear. At 8 months' follow-up, the surgical defect had healed completely and the patient has had no recurrent drainage or headache.

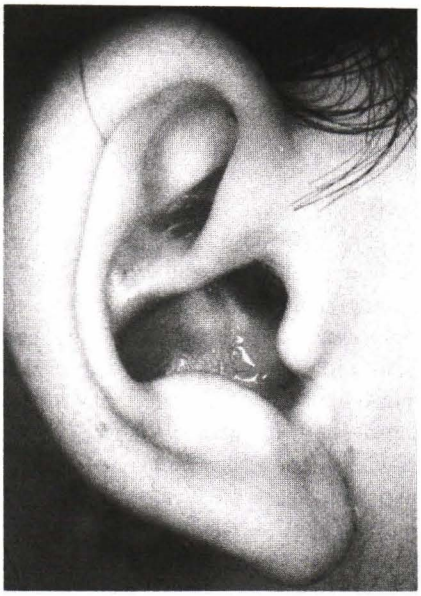

Fig. 1. Demonstrates right postauricular fistula and discharge from the right ear.

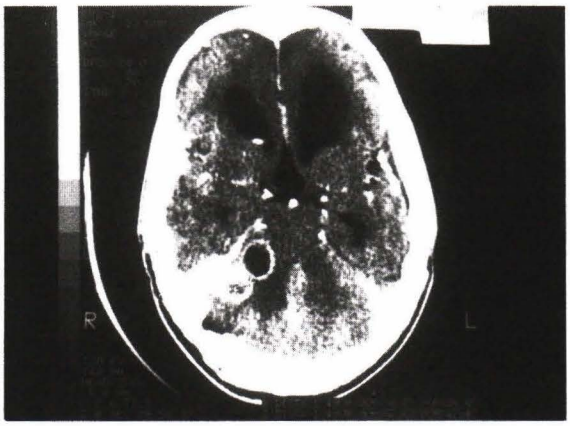

Fig. 2. Computed tomography of brain showing a brain abscess at the right cerebellar hemisphere.

Case 2

A 43-year-old male first presented at the age of 14 years when he had fractures of both tibias after a minor trauma. Later he developed sinusitis and oroantral fistula of the left maxillary sinus following a tooth extraction at the age of 31. Treatment included antibiotics, left Caldwell-Luc operation and closure of the oroantral fistula with a palatal flap. Post-operatively, he developed chronic osteomyelitis of the left maxillary antrum (Fig. 3). He was treated with parenteral antibiotics and curettage but the condition still persisted.

He came to follow-up regularly and was treated with multiple courses of intravenous antibiotics, as well as endoscopic removal of granulation tissue and sequestrum several times. At the age of 42, he developed a draining sinus tract through the skin 2.5 cm below the left lateral canthus. Exploratory surgery showed the antrum was filled with necrotic tissue and bony sequestra. A left partial maxillectomy was then performed and the sequestra removed. After the operation, he had no pain or drainage and successfully ware a maxillary denture with an obturator to fill the surgical defect. At 6 months follow-up, the surgical defect was filled with necrotic tissue and foul-smelling drainage. The patient was treated with intravenous antibiotics, curettage combined with hyperbaric oxygen. At 6 months'

follow-up, the surgical defect was nearly completed epithelized and the patient had a slight crust and drainage. The plan of treatment was to continue hyperbaric oxygen, as this treatment may can cure osteomyelitis.

DISCUSSION

The reported incidence of osteopetrosis ranges between 1:100,000 and 1:500,000[5]. Typical clinical findings are fragile bones, failure to thrive, hepatosplenomegaly, thrombocytopenia, and increased susceptibility to infection[4]. Patients usually die during infancy, but life expectancy has increased with improved medical care[6].

The characteristic pathophysiological features of osteopetrosis are the result of the lack of osteoclastic function. The osteoclast is a large multinucleated cell whose function is to absorb and remove osseous tissue. When the number and function of the osteoclasts decrease, "marbling" of the bone occurs which is characterized by sclerosis, brittleness, and radiopacity (Fig. 4)[7].

Osteomyelitis of the mandible is a clinical manifestation of osteopetrosis that is of great concern to the otolaryngologist. An increased incidence of osteomyelitis in the childhood form of osteopetrosis has been well established[3,8-10]. the authors reported one case of the childhood form (Case 1), who first presented with osteomyelitis of the mandible, bilateral optic atrophy and complete right facial palsy at the age of seven years. By 18, she had developed recurrent otitis media with a brain abscess. Because such a complication is severe and life-threatening, otolaryngologists need to make themselves aware whether a patient has underlying osteopetrosis which could develop into chronic otitis media. Chronic otitis media in patients with osteopetrosis is difficult to treat and the condition may progress to osteomyelitis of the temporal bone or develop intracranial complications such as brain abscesses. Osteopetrosis with otogenic brain abscess has not been reported in the literature. During the last 20 years, the mortality rate from brain abscess ranged from 13-40 per cent[11]. Early recognition of brain abscess with CT scan, appropriate systemic antibiotics and effective treatment of primary sources of infection, may reduce the mortality rate from brain abscess.

Case 2 had the adult form of osteopetrosis and was not as severe as the childhood form. He developed multiple fractures at 14 years of age. At 31, he developed sinusitis and an oroantral fistula following a tooth extraction. Because of the underlying osteopetrosis, the wound healed poorly and sinusitis progressed to osteomyelitis of the maxil-

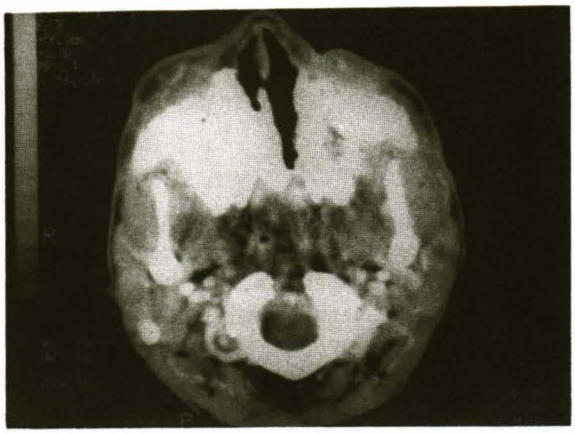

Fig. 3. Computed tomography of paranasal sinuses showing bony destruction of left maxillary sinus.

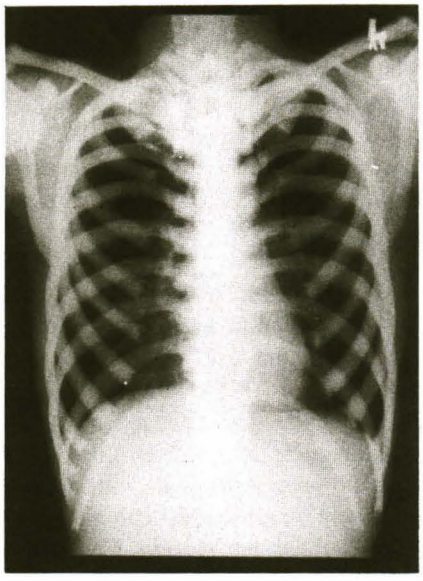

Fig. 4. Chest X-ray from case 2 demonstrating the typical radiologic appearance of osteopetrosis. Note the marbling of the bone and the dense bone formation.

lary bone. This development from tooth extraction must be a rare complication. Maxillary osteomyelitis secondary to osteopetrosis was successfully treated with a prolonged course of intravenous antibiotics and partial maxillectomy[12,13]. In the presented patient recurrent drainage was done 6 months after the initial treatment which may have been from severe extension of the disease. Hyperbaric oxygen has been reported as successful treatment of mandibular osteomyelitis in osteopetrosis[14]. In the presented patient the disease improved with hyperbaric oxygen and continued treatment is planned to cure the disease.

SUMMARY

Osteopetrosis is a rare disease and those afflicted with it are prone to serious complications. The authors encountered two cases of osteopetrosis with otolaryngological complications. The first case had chronic otitis media with brain abscess and the other had chronic osteomyelitis of the maxillary bone from tooth extraction.

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ภาวะแทรกซ้อนทางหู คอ จมูก ของโรคออสตีโอพีโทรสิส

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ออสตีโอพีโทรสิส เป็นโรคทางกระดูกที่ถ่ายทอดทางพันธุกรรมและพบได้น้อย พบได้ทั้งในมนุษย์และสัตว์เลี้ยงลูกด้วยนมอื่น ๆ ผู้เขียนได้รายงานคนไข้สองรายที่เป็นโรคนี้และมีภาวะแทรกซ้อนทางหู คอ จมูก คนแรกเป็นโรคนี้ชนิดในเด็กซึ่งมีความรุนแรงของโรคมากกว่าชนิดผู้ใหญ่ โดยมีน้ำหนวกไหลจากหูเรื้อรังและมีฝีในสมองร่วมด้วย ส่วนอีกรายหนึ่งเป็นผู้ใหญ่มีอาการไซนัสอักเสบภายหลังจากไปถอนฟันซึ่ง ต่อมาได้ลุกลามเป็นกระดูกอักเสบของกระดูกไซนัสบริเวณหน้า

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