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Modern Concept In Malignant External Otitis

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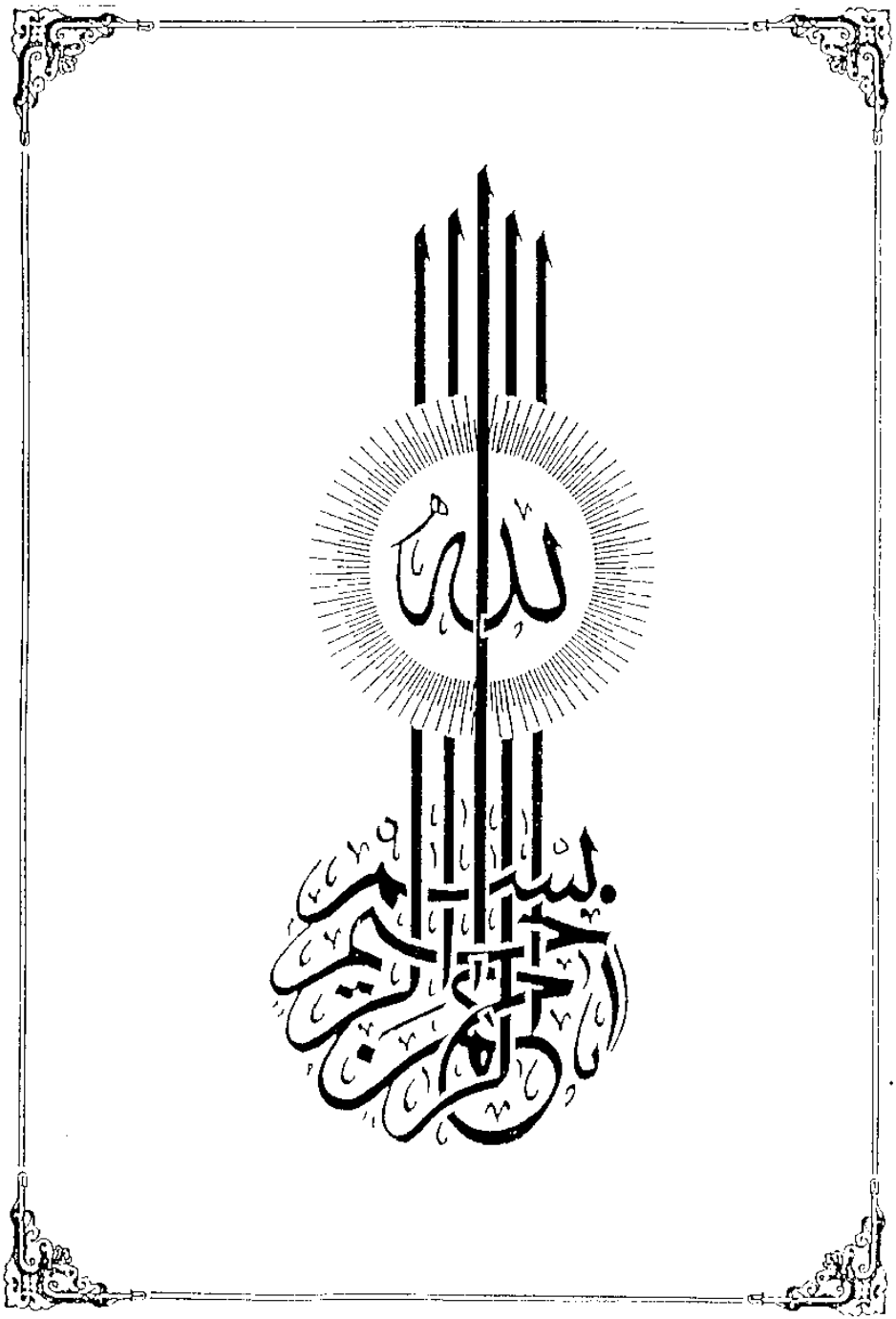

C O N T E N T S

INTRODUCTION

Simple otitis externa is a generally benign, self limited infection at the external ear canal seen in otherwise healthy individuals. (Sherman et al., 1980)

Recently, attention has been drawn to another variety of otitis which is unique, not only by virtue of its site of origin but also by its pattern of neurologic complications and the way in which these complications are produced. This condition, called"malignant external otitis", is an invasive bacterial infection of the external auditory canal with high morbidity and a 20% mortality rate despite appropriate antibiotic therapy (Rubin et al., 1990).

It is a disease predominantly of elderly diabetics caused mainly by Pseudomonas aeruginose that frequently progresses to an osteomyelitis of the base of the skull. Entry into the soft tissues of the parotid space takes place at the bony-cartilaginous junction of the external canal, accompanied by cellulitis at the stylomastoid foramen, which may cause facial nerve paralysis (Shupak et al., 1989).

Although it usually occurs in elderly, poorly controlled diabetic patients, cases do present in the middle aged and even

children and infants without history of diabetes. So, pediatricans should be aware that this serious infection can occur in young age without diabetes (Sherman et al., 1980).

The management of patients shown to have malignant external otitis should include strict diabetic control, regular local aural toilet and a six weeks course of parenteral and local antipseudomonal antibiotic agents. Together with metronidazole to cover any anaerobic element in the infection. The aim of this regimen is to reduce the morbidity and mortality of patients suffering from malignant external otitis. (Morrison et al., 1988).

AIM OF THE WORK:

The aim of this work is to review the literature dealing with this serious disease, its presentation, complications, evaluation and management.

DEFINITION

Toulmouche was the first to describe a case of malignant otitis externa with progressive osteomyelitis of the temporal bone in the Medical Gazette of Paris in 1938. In 1959, Meltzer and Kelman described a case of progressive pseudomonas osteomelitis of the temporal bone, zygoma, jaw and skull base.

Chandler, however, was the first to coin the term 'malignant external otitis', in 1968. It starts by pain and swelling of the external auditory meatus (this is why it was called external otitis). Soon paralysis of the last 4 cranial nerves appear and a fatal outcome occurs in many cases. This is why it has been called "malignant external otitis" of course not meaning malignant tumour, but having a malignant course (Badrawy, 1975).

Because the process is not neoplastic but represents an external otitis that invades adjacent structure, the term "invasive external otitis" was suggested to describe accurately the disease process that begins as a locally invasive Pseudomonas infection in the external ear canal, penetrates the epithelial barrier and results in signs of subcutaneous tissue invasion (Doroghazi et al., 1981).

The infection is characterized by the propensity of Pseudomonas aeruginosa to invade blood vessels and spread through soft and hard tissues causing vast necrosis and sc, proposed the term "necrotizing external otitis". (Rubinstein et al., 1980).

Many authors stress the need for this disease to be described as "necrotizing" instead of "malignant" otitis externa, as these term can lead to confusion with a neoplastic process. The similarity of symptoms between necrotizing otitis externa and carcinoma of the external auditory canal makes this semantic clarification even more necessary. (Mattucci et al., 1986, Sabater et al., 1988, Shupak et al., 1989).

As originally described by Chander in 1968, malignant external otitis is an infection of the external ear canal, usually due to Pseudomonas aeruginosa, which is associated with systemic invasion, significant neurologic sequelae and a high mortality rate. The vast majority of cases have occurred in elderly adults with diabetes mellitus. (Sherman et al., 1980).

Babiatzki and Sadè (1987) defined malignant external otitis as following:

1. A clinically severe external otitis which does not improve after 8 days of conservative treatment.
2. Pain, especially at night, which worsens after this period.
3. The presence of granulation tissue mostly over the floor of the external canal.
4. A culture of Pseudomonas pyocyaneus, usually.
5. A positive temporal bone scan, sooner or later.
6. Diabetes, in most patients.

Kimmelman et al., (198C) also defined malignant external otitis as a progressive, destructive osteomyelitis of the skull base usually originating in the region of the bony external auditory canal. auditory canal. The infection probably begins as a typical external otitis due to Pseudomonas aeruginosa, which progresses to osseous involvement in individuals with abnormal leukocyte function, such as elderly diabetics.

APPLIED ANATOMY OF THE EXTERNAL EAR

The ear is divided into three parts, the external ear, the middle ear and the inner ear.

The external ear is the part external to the tympanic membrane. It consists of the auricle and the external auditory canal (Chakeres et al., 1985). The external auditory canal extends from the concha of the auricle to the tympanic membrane. This distance is approximately 2.5 cm.

The inner two thirds of the canal consists of thin squamous epithelium and subcutanecus tissue applied directly to the periosteum. Although the tympanic bone makes up the greater part of the canal, and also carries the sulcus, the squamous bone forms the roof.

While the outer one third is supported by cartilage continuous with the auricular cartilage and is deficient superiorly, this space being occupied by the intrinsic ligament between the helix and tragus. The medial border of the meatal cartilage is attached to the rim of the bony canal by fibrous bands. The skin overlying the cartilaginous portion contains hairs and glands. The glands are of two types, ceruminous and sebaceous. Santorini's fissures pass through the cartilagenous floor of

the external auditory canal (Caruso et al., 1977) filled with nerves, vessels and connective tissue (Chandler, 1968).

The facial nerve exits from the stylomastoid foramen near the floor of the external auditory canal.

The jugular foramen, carrying the 9th, 10th and 11th cranial nerves, the hypoglossal canal carrying the 12th cranial nerve and the foramen oval carrying the mandibular branch of the trigeminal nerve are lying on the base of the skull within reach of an infection starting in the external auditory canal.

The parotid gland and the tempromandibular joint lie anterior and adjacent to the external auditory canal.

The medial relation to the external auditory canal are the tympanic membrane, the middle ear and ossicles.

The mastoid air cells system may be infected by ascending infection from the middle ear or by invasion of the bone of the mastoid cortex overlying the mastoid cells.

Mastoiditis may extend from the mastoid air cell system to the petrous apex reaching the middle or the posterior cranial fossa (Caruso et al., 1977).

A E T I O L O G Y

Malignant external otitis is more prevalent on repeated exposure to heat and moisture such as in humid climates or with frequent swimming (Dinapoli, 1971). So it is more prevalent in the summer when external otitis is rampant (Babiatzki and Sadè, 1987). This leads to decrease in the production of cerumen, which is bacteriostatic and funqistatic, and surface infection develops (Dinapoli, 1971).

In some years, a relatively large number of these patients appear, in others there are none. The disease is more prevalent in Jews than in Arabs. The reason for this is unknown (Babiatzki and Sadè, 1987).

Predisposing Factors:

Diabetes mellitus has been associated with a number of infectious complications and malignant external otitis occurs primarily (93% of cases) in diabetic patients (Zaky, 1976). Why is the diabetic patient's ability to respond to infections impaired?

Lucente et al., (1982) stated that there are several reasons. First, diabetics demonstrate poor leukocyte migration and decreased cellularity at inflammatory sites, reflecting impairment of the cellular defense mechanism. Secondly,

defective phagocytosis occurs both during Ketoacidosis and nonketotic stages. And thirdly, the diabetic shows reduced lymphocyte reactivity phytohemagglutinin stimulations and evidence of depressed antibacterial defenses in the presence of a high concentration of serum glucose.

Also, Resouly et al., (1982) stated that the pathogenesis of malignant external otitis depends on apportunistic infection by Pseudomonas aeruginosa in avascular bone and diabetics are especially at risk as a result of poor vascular supply and impaired phagocytic defence mechanism in the presence of hyperglycaemia.

The polymorphonuclear leukocyte system, an intergral part of the host defence mechanism against Pseudomonas, is defective in the diabetic patients. Similary, these patients show impairment in the production of an opsonizing antibody that is intergral to this protection system (Ostfeld et al., 1981).

Ostfeld and associates (1981) have recently suggested an another reason for the peculiar susceptibility of the diabetic patients. They have demonstrated a thick layer of hypocellular degenerated collagen or perichondrium extending from the cartilage into the dermis in two patients and have speculated

that the disease results from invasion of apportunistic organisms into previously devitalized tissues.

The combination of diabetes with advanced age probably cause weakening of the natural defense mechanisms possibly created suitable conditions for Pseudomonas infection to become malignant (Joachims, 1976).

Shupake et al., (1989) reported two cases, both were elderly insulin dependent diabetics and the patient's record included cerebral ischemic attack and peripheral arterial vascular disease. So, patients with history of underlying peripheral vascular diseases and elderly diabetics, named the group most prone to develop malignant external otitis.

More recently, Rubin et al., (1990) suggested that elderly patients with diabetes mellitus, the high risk group for acquiring this serious disease, have a diminished cutaneous barrier against infection because of microangiopathy associated with diabetes.

The few malignant external otitis patients reported who were not diabetics suffered from diseases associated with immunosuppression, such as Leukemia and granulocytopenia (Chandler., 1977; Meyrhoff et al., 1977).

While the disease was originally described in elderly diabetics, and diabetes and old age are still regarded as the most important risk factors in adult, review of the pediatric case reports stresses anaemia poor general condition and concurrent systemic disease as predisposing factors for malignant external otitis in children (Joachims et al.,1990).

Two cases reported by Joachims (1976), prior to the onset of ear infection, they suffered from serious systemic diseases; one from bronchopneumonia and the second from gastroenteritis.

Also, three pediatric case were immune depressed - one due to cytotoxic treatment for neuroblastoma (Rubinstein et al., 1980) another due to intoxication induced aplastic anaemia (Schimmel et al., 1980) and the third due to cyclic neutropenia (Ichimura et al., 1983).

Two previously healthy children acquired malignant external otitis during an episode of Stevens-Johnson syndrome (Sherman et al., 1980); Horn and Gherini, 1981).

Thus, weakening of natural defense mechanisms possibly created suitable conditions for Pseudomonas infection to become malignant (Joachims, 1976).