The Journal of Laryngology and Otology
June 1990, Vol. **104**, pp. 488–490

Clinical Records

Malignant external otitis in an infant

DAN NIR, M.D.*, TSILA NIR, M.D.†, JOSHUA DANINO, M.D.* and HENRY Z. JOACHIMS, M.D.*

Abstract
Ten cases of malignant external otitis in children have been reported hitherto. These are reviewed, and an eleventh case, a three-month-old infant associated with genetic granulocytopenia, is presented. On the grounds of the reported paediatric cases, the erudition in childhood is compared with the more common form in the adult, and is found to be much less linked with diabetes mellitus and to have a far better prognosis, with practically no mortality.

Introduction

The term 'malignant external otitis' (MEO) was first coined by Chandler in 1968 to describe a particularly severe form of infection differing from both diffuse and localized external otitis by its progressive course and poor response to treatment. The disease is characterized by unremitting pain and purulent discharge, and it tends to invade cartilage, bone, nerve and adjacent soft tissues. It affects primarily elderly diabetics, and the causative agent is almost uniformly *Pseudomonas aeruginosa*. It may progress to involve the base of skull with multiple cranial nerve palsies and meningitis, leading to death in 30 to 80 per cent of patients (Chandler, 1977; Damiani *et al.*, 1979; Doroghazi *et al.*, 1981; Mills, 1986).

Since Chandler's original description, cases with MEO have been reported in non-diabetic debilitated patients (John and Hopkins, 1978), in patients on cytotoxic chemotherapy (Rubinstein *et al.*, 1980) and in young diabetics (Merritt *et al.*, 1980; Shamboul and Burns, 1983).

The first report of paediatric MEO in the English literature was by Joachims (1976) who described two non-diabetic infants, aged 7 months and two years; both were complicated by facial paralysis. These first two cases were followed in the same year by Giguere and Rouillard's report (1976) of a 10-year-old diabetic girl with malignant external otitis and facial nerve palsy. Further case reports in children are those of Coser *et al.* (1980) (aged 5 and 6 months); Schimmel *et al.* (1980) (age 10 years); Rubinstein *et al.* (1980) (age 4 months); Sherman *et al.* (1980) (aged 6 years), Horn and Gherini (1981) (aged 6 years) and Ichimura *et al.* (1983) (aged 2 years). Doroghazi *et al.* (1981), in a review of 21 adult patients with malignant external otitis, mention their youngest patient to be an 11-year-old non-diabetic male. Altogether, only ten cases of MEO have been described in the paediatric age group, four of these were anaemic and in poor general health (Joachims 1976; Coser *et al.*, 1980), one was a juvenile diabetic (Giguere and Rouillard, 1976), three 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 were previously healthy but acquired the disease during an episode of Stevens-Johnson's syndrome (Sherman *et al.*, 1980; Horn and Gherini, 1981). No further information was obtainable on Doroghazi's patient (1980) except for the fact that he was non-diabetic and was cured of disease.

We wish to present an eleventh case, known by her family history and laboratory examination to have Genetic Agranulocytosis, who presented to us with malignant external otitis. Infantile Genetic Agranulocytosis is an autosomal recessive disease, first described by Kostman in 1956. It is characterized by frequent episodes of infection commencing early in life, with extremely low blood neutrophil counts due to bone marrow maturation abnormalities. The severe neutropenia of this disorder usually leads to fatal infection (Williams *et al.*, 1983).

Case report

A 3-month-old female infant, third offspring to young, non-consanguineous parents of Arab descent, was first seen by her family physician due to restlessness and right aural discharge. With the diagnosis of acute otitis media, oral ampicillin was prescribed. After nine days of treatment with no apparent improvement and a rise in temperature up to 39°C, the patient was referred to our paediatric emergency ward. The child was born weighing 3960 g, after a normal pregnancy and delivery. Her eldest brother suffered from recurrent infections, was diagnosed as having Kostman's disease, and succumbed to it at 3½ years of age. A second brother, now 6 years old, suffers frequent episodes of severe infection, usually in the region of the orbits and oral cavity, and granulocytopenia.

Routine blood count of our patient one month prior to her illness revealed a WBC of 10,000, with 1% polymorphs, and she too was diagnosed as having Kostman's disease. Physical examination on admission revealed a well nourished, slightly pale, extremely restless infant. The right ear was draining with copious purulent discharge, with erythema and swelling of the skin of the external meatus, tragus and pretragal area. The skin of the external canal was swollen and inflamed, and a mass of hard white tissue (later found to be denuded cartilage) partially occluded the lumen. The eardrum could not be seen. There was no facial weakness, nor was any other cranial neuropathy observed. The left ear and the rest of the examination were unremarkable. Fever was 38.5°C, ESR—112/h, Hb—8.3 g/dl and WBC 13,500/cc with 2% polymorphs, 67% lymphocytes, 30% monocytes and 1% eosinophils. Cultures of the aural debris grew *Pseudomonas aeruginosa* and *Proteus rettgeri* and,

From the Departments of *Otolaryngology and †Paediatrics B, Rambam Medical Center and Faculty of Medicine, Technion-Israel Institute of Technology, Haifa, Israel.
Accepted for publication: 23 February 1990.

https://doi.org/10.1017/S0022215100112952 Published online by Cambridge University Press

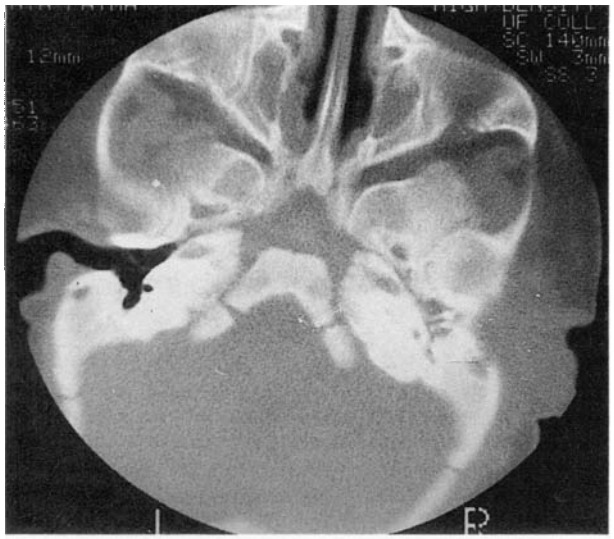


FIG. 1

CT scan demonstrating swelling of soft tissues in the right external canal and partial erosion of its posterior wall.

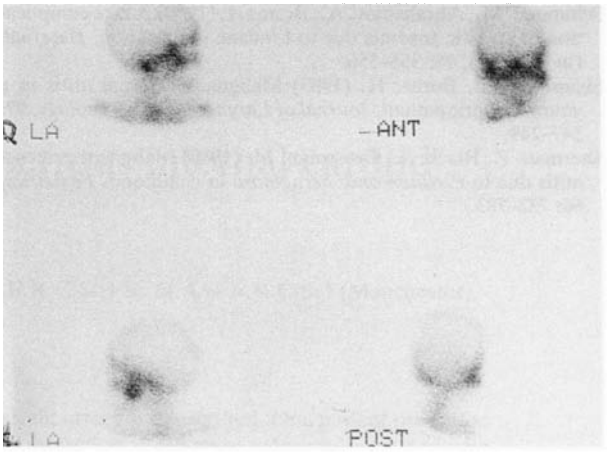


FIG. 2

Gallium scan demonstrating increased uptake in the area of the right external ear.

according to sensitivity, treatment was begun with intravenous gentamicin and cefotaxime. The fever subsided 48 hours later but aural discharge continued despite local cleaning and application of ear drops. On the ninth day, under general anaesthesia, the hard white mass in the inflamed external canal was excised and histologically diagnosed as inflamed cartilage with areas of necrosis. The ear drum was normal. The diagnosis of malignant external otitis was made, and respectively treatment was begun with intravenous amikacin and piperacillin. Surgical debridement and removal of granulation tissue and necrotic cartilage was performed shortly thereafter.

Computed tomography demonstrated severe swelling of soft tissues in the right external canal, extending anteriorly to the parotid gland and posteriorly over the mastoid. The posterior wall of the bony canal was partly eroded (Fig. 1). The structures of the middle and inner ear were normal. In a Tc scan of the right temporal bone there was no pathological uptake, yet Ga uptake was localized to the area of the external canal, suggesting an inflammatory process of the soft tissues (Fig. 2). Repeated cultures from the aural discharge revealed *Pseudomonas aeruginosa* and *Proteus rettgeri* at first, Pseudomonas only later, and by the fourth week of treatment cultures became sterile. Blood cultures were sterile all along the disease. A hydrocortisone-rafusone-soaked pack was put in the ear canal and changed every other day, in an attempt to minimize external canal stricture. The discharge gradually ceased and the patient was discharged after completing six weeks of therapy. Renal function was monitored weekly during the treatment period and remained normal. The patient was closely observed twice weekly in the outpatient clinic for a period of three months, and later once a month. To date, 20 months following the event, the patient is free of aural disease. She has since been hospitalized twice—once for acute gastroenteritis and a second time for osteomyelitis of the femur. Hearing evaluation on follow-up was normal.

Discussion

In nearly 20 years since Chandler's first description of a necrotizing infectious process of the external ear which he termed 'malignant external otitis', only 10 cases were reported in the paediatric age group. While the disease was originally described in elderly diabetics, and diabetes and old age are still regarded as the most important risk factors in adults, review of the paediatric case reports stresses anaemia, poor general condition, and concurrent systemic disease as predisposing factors. Only one of the reported children was a juvenile diabetic (Giguere and Rouillard, 1976), compared with 89% in adults (Doroghazi *et al.*, 1981). Another major difference between paediatric malignant external otitis and the adult disease is its outcome. Though mortality in adults is still very high, all but one of the 11 reported children were totally cured from the disease. The only exception was a 10-year-old boy who was in an almost terminal state prior to acquiring the MEO (Schimmel *et al.*, 1980). The patient was hospitalized because of aplastic anaemia following ingestion of Lindane (gamma benzene hydrochloride), with repeated septicaemia and pneumonia, and after three months of unsuccessful treatment developed MEO, of which he died two weeks later. Our patient, though suffering from an immunological defect, fared well with adequate antibiotic treatment and surgical debridement, and was completely cured of the disease.

References

Chandler, J. R. (1968) Malignant external otitis. *Laryngoscope,* **78:** 1257–1293.

Chandler, J. R. (1977) Malignant external otitis: further considerations. *Annals of Otology, Rhinology and Laryngology,* **86:** 417–428.

Cóser, P. L., Stamm, A. E. C., Lobo, R. C., Pinto, J. A. (1980) Malignant external otitis in infants. *Laryngoscope,* **90:** 312–316.

Damiani, J. M., Damiani, K. K. and Kinney, S. E. (1979) Malignant external otitis with multiple cranial nerve involvement. *American Journal of Otology,* **1:** 115–120.

Doroghazi, R. M., Nadol, J. B. Jr, Hyslop, N. E. Jr, Baker, A. S., Axelrod, L. (1981) Invasive external otitis: report of 21 cases and review of the literature. *American Journal of Medicine,* **71:** 603–614.

Giguere, P., Rouillard, G. (1976) Otite externe maligne bilaterale chez une fillette de 10 ans. *Journal of Otolaryngology,* **5:** 158–166.

Horn, K. L., Gherini, S. (1981) Malignant external otitis of childhood. *American Journal of Otology,* **2:** 402–404.

Ichimura, K., Hoshino, T., Yano, J., Nozue, M. (1983) Neutrophil disorders in a child with necrotizing external otitis. *Journal of Otolaryngology,* **12:** 129–133.

Joachims, H. Z. (1976) Malignant external otitis in children. *Archives of Otolaryngology,* **102:** 236–237.

John, A. C., Hopkins, H. B. (1978) An unusual case of necrotizing otitis externa. *Journal of Laryngology and Otology,* **92:** 811–812.

Merritt, W. T., Bass, J. W., Bruhn, F. W. (1980) Malignant external otitis in an adolescent with diabetes. *Journal of Paediatrics,* **96:** 872–873.

Mills, R. (1986) Malignant otitis externa (Editorial). *British Medical Journal,* **292:** 429–430.

Rubinstein, E., Ostfeld, E., Ben-Zaray, S., Schiby, G. (1980) Necrotizing external otitis. *Pediatrics,* **66:** 618–620.

https://doi.org/10.1017/S0022215100112952 Published online by Cambridge University Press

Schimmel, M., Abrahamov, A., Brama, I. (1980) A rare complication of aplastic anaemia due to Lindane intoxication. *Harefuah* (in Hebrew), **98:** 355–356.

Shamboul, L., Burns, H. (1983) Malignant external otitis in a young diabetic patient. *Journal of Laryngology and Otology,* **97:** 247–249.

Sherman, P., Black, S., Grossman, M. (1980) Malignant external otitis due to *Pseudomonas aeruginosa* in childhood. *Pediatrics,* **66:** 782–783.

Williams, W. J., Bentler, E., Erslev, A. J., Lichtman, R. W. (1983) *Hematology,* 3rd ed. New York, McGraw-Hill, pp 778–779.

Address for correspondence:
Dan Nir, M.D.
Department of Otolaryngology
Rambam Medical Center
POB 9602, Haifa 31096, Israel

https://doi.org/10.1017/S0022215100112952 Published online by Cambridge University Press