Case Report

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Malignant otitis externa in a 21-year -old male patient with Prader–Willi syndrome

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Abstract

Malignant otitis externa is an invasive infection of the external auditory canal and temporal bone with potentially life-threatening complications. Elderly patients with type 2 diabetes mellitus are the population most commonly affected by malignant otitis externa, but any type of immunosuppression predisposes to the disease. Prader–Willi syndrome is a genetic cause of obesity, often associated with insulin resistance and type 2 diabetes mellitus. This report describes a case of a 21-year-old male patient with Prader–Willi syndrome who had malignant otitis externa that progressed to sepsis during hospitalization. To the best of the authors' knowledge, this is the first description of malignant otitis externa in a young patient with Prader–Willi syndrome.

Keywords

Otolaryngology, obesity, malignant otitis externa, Prader–Willi syndrome

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Introduction

Malignant (necrotizing) otitis externa (MOE) is a type of temporal or base of the skull bone osteomyelitis. Generally, it is a complication of an initial diffuse otitis externa that has potentially life-threatening complications. The most susceptible patients are those with type 2 diabetes mellitus (DM2), but any type of immunosuppression predisposes to the disease. The most frequent pathogens are those related to the mucus-cutaneous microbiota and *Pseudomonas aeruginosa* is the most common one.[1] Other described pathogens are methicillin-resistant *Staphylococcus aureus* (MRSA), *Enterobacter, Klebsiella pneumoniae*, and Proteus.[2] Initially, patients complain of otalgia—more severe at night—and otorrhea. In advanced stages, excruciating pain with low response to common painkillers and purulent foul-smelling otorrhea are described. Cranial nerve involvement is quite common and may be present in as much as 43.5% of all cases.[3]

The authors report the case of a 21-year-old male patient with Prader–Willi syndrome (PWS), who had MOE that progressed to sepsis during hospitalization. All procedures performed were in accordance with the ethical standards of the 1964 Helsinki declaration and its later amendments. After extensive review of the literature, the authors believe that this is the first description of MOE in a young patient with PWS.

Case report

A 21-year-old male patient with diabetes, hypertension, and PWS presented to the Emergency Department with otalgia, otorrhea, and fever for 2 weeks without improvement following treatment regimes including amoxicillin–clavulanate, ciprofloxacin, and ceftriaxone.

On arrival, he was tachypneoic with increased respiratory effort and had significant left-sided otorrhea; laboratory tests indicated diabetic ketoacidosis. Computed tomography (CT) scans of the head and temporal bones revealed small areas of mastoid cell erosion on the left side; significant discharge with some gas focuses within the external ear canal, tympanic cavity, antrum, and left mastoid cells; these changes extended to the extracranial soft tissues, left face, and cervical region, reaching the skin, the external ear, and the parotid

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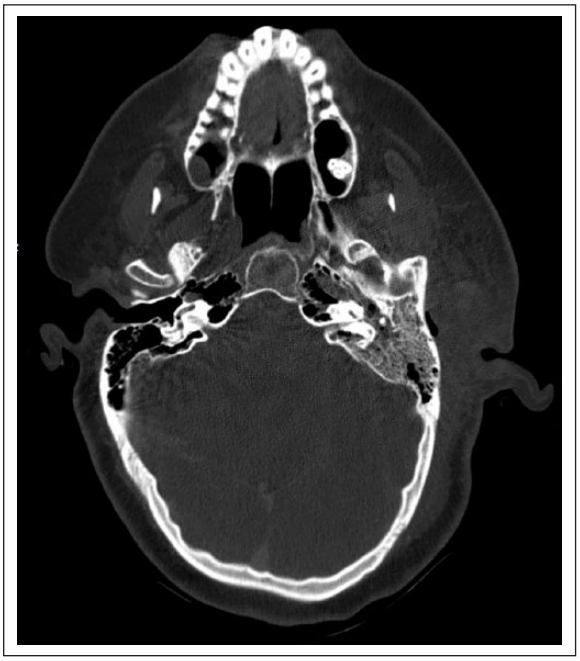

Figure 1. Mastoid and cranium computerized tomography at hospital admission with small areas of probable mastoid cells discontinuity on the left side and important amounts of discharge with some sparse gas focuses.

gland (Figure 1). Lumbar puncture excluded the spread of infection to the central nervous system.

Initially, in addition to necrotizing otitis media, parotid abscess was also considered as a diagnosis because of the tomographic features. Hence, the first antimicrobial regimen chosen was vancomycin associated with meropenem in order to cover MRSA and *Pseudomonas*, as recommended by the hospital infection control committee. Due to worsening of the respiratory pattern and the development of metabolic acidosis at the second day of the hospital stay, he was transferred to the ICU, requiring orotracheal intubation and mechanical ventilation for 6 days. On the fifth day of antibiotic therapy, due to the sustained absence of fever and the evidence of low serum level of vancomycin (12 mcg/mL), it was considered that the MRSA coverage was no longer needed. Thus, vancomycin was discontinued and the regimen was changed to meropenem plus ciprofloxacin and dexamethasone combination ear drops, which was maintained until discharge.

After being extubated and discharged from the ICU, the patient developed fever, productive cough with mucopurulent sputum, and worsening of the ventilatory pattern, with need for noninvasive ventilation. Polymyxin B was added for 8 days to treat hospital acquired pneumonia, progressing with improvement of the ventilatory pattern and absence of fever.

Technetium-99m medronic acid (^{99m}Tc-MDP) scintigraphy performed at the 21th day of hospitalization revealed an abnormal high uptake area in the mastoid process of the left temporal bone. Galium-67 scintigraphy performed at the same day revealed the same findings.

Improvements of ear and motor symptoms as well as clinical and laboratory tests were observed after 36 days of hospitalization. The patient was discharged in good condition with the prescription of ciprofloxacin 750 mg 12/12 h and outpatient care.

A new Galium-67 bone scintigraphy done 30 days after discharge showed very significant decrease of the active infection/inflammation at the left temporal region. Antibiotics were maintained. Three months after discharge, another Galium-67 scintigraphy did not show any abnormality in the area where previously there was increased uptake of the radiation marker due to the MOE (Figure 2). Thus, antibiotic treatment was discontinued.

Discussion

MOE is an infection of the external ear canal and the skull base caused by *P. aeruginosa* in more than 90% of the cases.[1] The diagnostic criteria for MOE proposed by Cohen and Friedman[4] include obligatory criteria (pain, exudate, edema, granulations on otoscopy, microabscess when operated, positive Technetium-99 uptake or failure of treatment more than 1 week, and possibly pseudomonas in culture) and occasional criteria (diabetes, cranial nerve involvement, positive radiograph, comorbidities, and old age). More recently, Joshua et al.[5] revised the diagnostic criteria for MOE, highlighting the fact that MOE may be present even when one of the major criteria is not met.[5] All but one obligatory criteria were present in this patient, who also presented occasional criteria (diabetes mellitus); unfortunately, ear discharge culture was not performed, and the result of blood cultures was absence of growth. With progression of the disease, temporal bone osteomyelitis associated with facial nerve palsy and extension to the central nervous system may occur, with subsequent meningitis, brain abscess, or thrombosis of the intracerebral venous sinuses.[6]

An analysis of 8300 inpatients with MOE recently published showed that the mean age of these patients was 54.1 years ($\pm$20.4). In addition, 55.1% had diabetes mellitus and 12.5% were obese. The mean hospital stay was of 4.8 ± 5.1 days, and sepsis occurred in 1.1% of the cases.[6] Among the special features of our patient are the early age of occurrence and the severity of the disease, as, unlike most cases, he had sepsis that caused a long hospital stay.

In a recent retrospective study carried out in South Korea, the morbimortality of MOE was significantly higher in patients with long-standing DM2, higher levels of serum inflammatory markers at presentation or jugular foramen and petrous apex involvement; infectious agents were not relevant as a prognostic factor. Concerning therapy options, there was no significant difference in terms of treatment outcomes between antibiotics alone, antibiotics

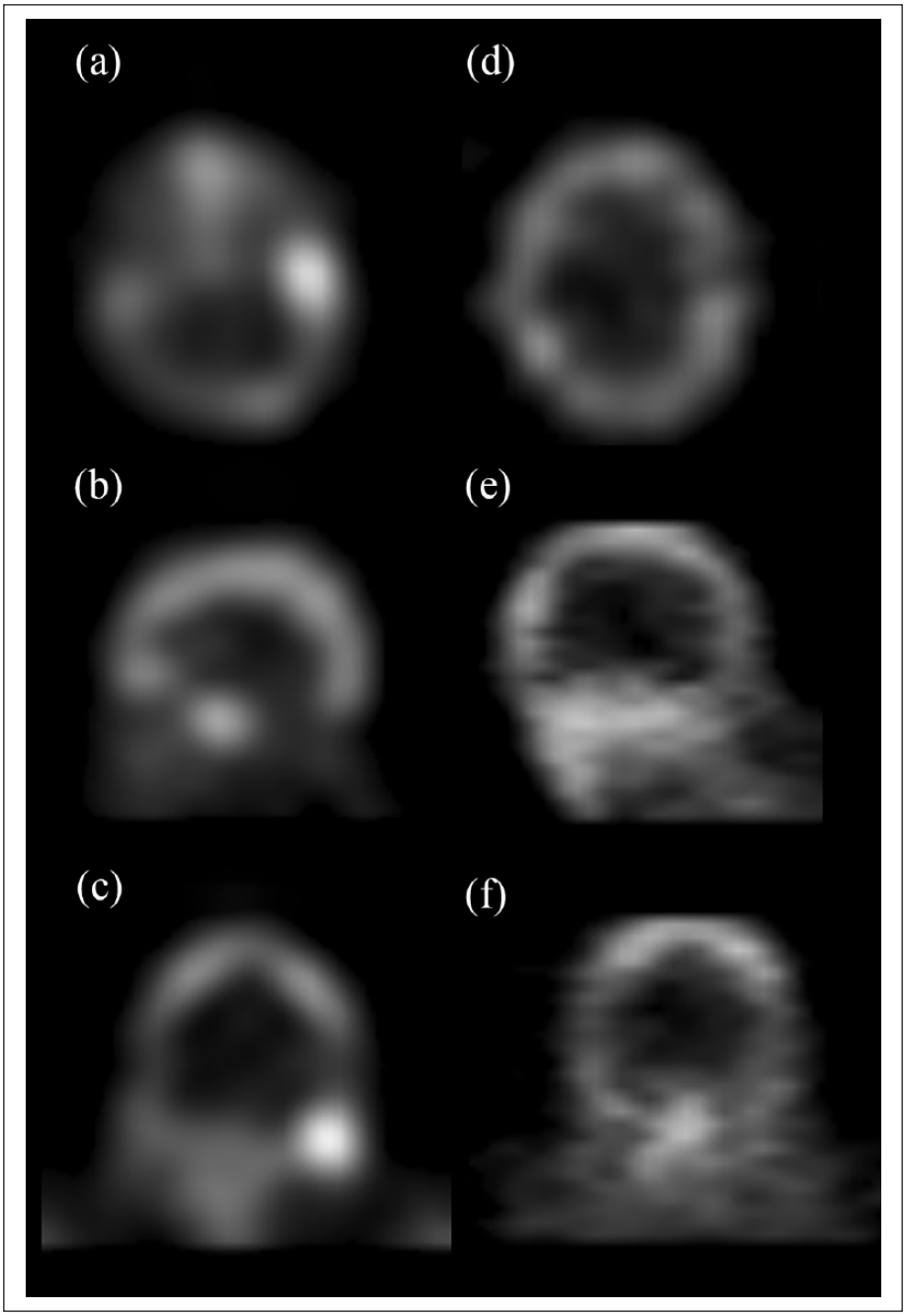

Figure 2. Galium-67 scintigraphy. Transversal (a), sagittal (b), and coronal (c) view at the 21th day of hospitalization, showing an abnormal high uptake area in the mastoid process of the left temporal bone. Transversal (d), sagittal (e), and coronal (f) view, without increased uptake of the radiation marker, 3 months after discharge.

in combination with steroids, or antibiotics plus surgery.[2] The authors of the present case report decided to use only antibiotics; initially, 4 days of vancomycin combined with meropenem and, thereafter, meropenem plus ciprofloxacin and dexamethasone combination ear drops.

Regarding diagnosis, CT scan of the temporal bone with contrast is appropriate and commonly used for initial assessment, being useful to exclude other ear pathologies. In MOE, it shows erosion of the tympanic bone and the skull base as well as the involvement of adjacent tissues. Magnetic resonance imaging might be performed in order to complement CT, once it is useful for better detailing of the disease spread and soft tissue involvement. Combined Technetium-99 and Gallium-67 scintigraphy are broadly used to define bone involvement, being Gallium-67 imaging especially useful for evaluating treatment response (follow-up).[7] When the patient of this case report arrived, the contrast CT scan showed an inflammatory/infectious process that involved the ear

structures, parotid gland, and adjacent cellular subcutaneous tissue. The Technetium-99 scintigraphy performed 20 days later showed uptake consistent with left ear MOE. Gallium-67 bone scintigraphy later performed corroborated the hypothesis of MOE.

The PWS is the most common genetic cause of obesity.[8] The most frequent complications among the patients with this syndrome are insulin resistance, atherosclerosis, and sleep apnea.[9] DM2 is another common finding and is one of the main risk factors for MOE as well. In spite of the apparent relation of these diseases, the authors did not find any literature on MOE involving young patients with PWS. Among other features of PWS, this patient has sleep apnea, hypogonadism, and behavioral problems.[9] Physical examination revealed infantile penis and severe phimosis. According to family members, he has emotional incontinence and lability that contribute to childish behavior. Despite his obvious behavioral problems, he has normal intelligence, inferred by the medical team, without the use of scales.

In conclusion, this is the first reported case of MOE, a typical affection of the elderly, in a 21-year-old patient with PWS, a genetic syndrome associated with obesity and DM2. This case highlights the importance of suspecting of MOE even in young patients, especially if risk factors for the disease are present, once early diagnosis can avoid or minimize life-threatening complications.

Declaration of conflicting interests

The author(s) declared no potential conflicts of interest with respect to the research, authorship, and/or publication of this article.

Ethical approval

Our institution does not require ethical approval for reporting individual cases or case series.

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Informed consent

Written informed consent was obtained from the patient(s) for their anonymized information to be published in this article.

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