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內文：

Summary：

Granulocytic sarcoma is an uncommon harbinger of myeloproliferative disorders. This case is the first report of a primary granulocytic sarcoma presenting with sole finding of an external auditory canal mass in an otherwise healthy newborn twin male. After an extensive immunohistochemical evaluation, the biopsy demonstrated monoblastic sarcoma, an uncommon subtype of granulocytic sarcoma more commonly seen in infants.

Introduction:

1. Granulocytic sarcomas (also called myeloblastomas or chloromas) are solid, extramedullary collections of myeloblasts occurring anywhere in the body.
2. This pathology is commonly associated with acute myelogenous leukemia (AML), and less typically with acute lymphocytic leukemia (ALL).
3. Granulocytic sarcomas are most commonly mistaken for non-Hodgkin's lymphoma (diffuse large cell type), soft tissue sarcomas, Ewing's sarcomas, or poorly differentiated epithelial tumors.

Case presentation：

1. A term fraternal twin male (twin A) was born via uneventful vaginal delivery. His hospital course was uncomplicated, including a normal hearing screen.
2. On DOL（day of life）5, the infant was noted to have right otorrhea which the parents regarded as cerumen(耳垢). On DOL 14 the infant was taken to an outside emergency department when the otorrhea was noted to be increasingly purulent and was treated with amoxicillin. Cultures returned Staphylococcus aureus.
3. The otorrhea persisted and, on DOL 21, his antibiotic regimen was changed to amoxicillin/clavulanic acid and ciprofloxacin/dexamethasone otic suspension.
4. Examination under anesthesia demonstrated a friable polypoid mass in the EAC. Due to neutropenia, a bone marrow aspirate was done to rule out congenital neutropenia. This demonstrated a normal bone marrow aspirate with sequential maturation and no evidence of malignancy.
5. The biopsy demonstrated atypical histiocytic infiltrate consistent with monoblastic sarcoma, M5 AML type. This was supported by immunohistochemical staining which was positive for lysozyme and CD68. Stains were negative for S-100, desmin, mast cell markers and tryptase.
6. He was treated with chemotherapy per Children's Oncology Group protocol AAML02P1, a pilot study for the treatment of children with AML.
7. He has been off therapy for 2 years. Bone marrow done at the end of treatment and serial CT scans showed no evidence of disease.

Discussion：

1. Primary granulocytic sarcomas or chloromas may be a harbinger of acute myelogenous leukemia, specifically, AML subtypes M1 and M2 in adults, and in

infants, M4 and M5.
2. Granulocytic sarcomas are associated with multiple chromosome abnormalities such as t (8;21) and inv (16) and rare monosomy, as well as focal expression of surface markers, CD 56, 2, 4, and 7.
3. When granulocytic sarcomas occur in bone, myeloblasts are thought to travel from the marrow through the haversian system to rest in the periostium, and there is a propensity for ligamentous involvement.
4. CD68 and lysozyme, both monocytic antigens, were positive yielding a diagnosis of monoblastic sarcoma.
5. They most commonly present in the orbit and nasopharyx. It is seen in approximately 3-5% of pediatric patients with acute myelogenous leukemia with a slightly lower incidence within the general population with equal prevalence in both sexes.
6. There is no role for surgery beyond biopsy, and rapid diagnosis is imperative to successful treatment as early chemotherapy has been successful in inducing complete remission.

Conclusion：

1. Granulocytic sarcomas should be suspected in cases of temporal bone tumors, even in the absence of systemic manifestations of leukemia at any age.
2. Recognition of this rare entity is important because early aggressive chemotherapy can bring about regression of the tumor and improve survival.

題號	**題目**
1	下列何者是對Granulocytic sarcomas較有效的治療方法？ **(A)** Radiation therapy **(B)** Chemotherapy **(C)** Surgery without biopsy **(D)** Antibiotics
答 案 (B)	出處：Oral & Maxillofacial PATHOLOGY (second edition) p.511、512、本篇JOURNAL
題號	題目
2	當我們懷疑是leukemia引起的Granulocytic sarcomas時,我們可以做下列檢測，何者有誤？ **(A)** Bone marrow biopsy **(B)** Cytogenic evaluation **(C)** Immunohistochemical markers for lysozyme **(D)** Immunohistochemical markers for CD-PRO-2
答 案 (D)	出處：Oral & Maxillofacial PATHOLOGY (second edition) p.512、本篇JOURNAL