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Asymptomatic palatine swelling with rapid worsening of young children

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1 **Title**

2 Asymptomatic palatine swelling with rapid worsening of young children

3

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22 **Manuscript**

23

24 **Observation**

25

26 A 2-year-old child without a medical history is referred to the paediatric emergency
27 department for advice regarding the aggravation of a right palatal swelling that
28 appeared 3 days earlier. In his letter, the attending physician reports the notion of a
29 suspicion of a primary varicella-zoster-virus infection dating back a week with the
30 initiation of a probabilistic antibiotic therapy with amoxicillin-clavulanic acid (1 dose
31 weight*3 per day) 72 hours ago, when this palatal mass was discovered, suggesting
32 serous dental cellulitis.

33 Clinically, the child presents a well-limited, painless right palatal mass that slightly
34 crosses the midline and respecting the palatal veil. This tumour is indurated, non-
35 renitent and non-inflammatory (Fig. 1). It presents no dietary difficulties but appears
36 very tired. No dental mobility or pain on percussion or cold test is found.

37 In view of the atypical history of the disease and the prospect of an injected imaging
38 exam to complete your observation, you decide to carry out a biological assessment
39 (blood count, CRP, kidney function, coagulation test). 30 minutes later, the biologist
40 who wishes to personally give you the results calls you urgently. The child presents
41 anemia at 7.1 g/dl (11<N<14), thrombocytopenia at 22,000 per mm³ (200<N<490),
42 hyperleukocytosis at 76,000 per mm³ (5,000<N<15,000) with a blood smear
43 reporting 48% myeloid blasts.

44

45 **What is your diagnosis ?**

46

47 **Response**

48

49 It is a Myeloid Sarcoma (MS) of the right maxilla (= solid tumour with extra-myeloid
50 cells), which in this observation is expressed in the context of acute myeloblastic
51 leukemia with non-hyper-leukocyte t(8;21) translocation.

52

53 **Discussion**

54

55 Myeloid sarcoma (MS) is a rare, localized tumour comprising immature granulocytic
56 cells. It is also called Chloroma because of its green colour induced by the
57 concentration of myeloperoxidase[1]. It may occur in pre-existing acute myeloid
58 leukaemia (AML) (3-8%), before the onset of AML (0.6%) or in association with
59 other myelo-proliferative conditions such as blast crisis in chronic myeloid leukaemia
60 and myelodysplastic Syndrome[2,3]. MS is primarily of soft tissue interest and
61 remains extremely rare in the head and neck area[4]. Nevertheless, its knowledge is
62 essential as it represents a real diagnostic and therapeutic emergency.

63 Confirmation of the diagnosis of MS is based on the performance of flow cytometer
64 of circulating blood and bone marrow, a myelogram, biopsy of the lesion and genetic
65 research since many mutations may be involved. Rapid determination of the
66 phenotype is crucial in order to best adapt the treatment protocol. The assessment
67 must be completed by an imaging examination (CT scan and/or magnetic resonance

68 imaging with injection of contrast agent) before the introduction of any therapy[2,5].
69 These examinations will allow the assessment of tumour anatomic ratios and will
70 serve as reference examinations for the evaluation of the efficacy of systemic
71 treatments. Indeed, the prognosis of MS depends essentially on the initial context in
72 which it is diagnosed (age, presence or not of AML)[5]. MS management is based on
73 the rapid introduction of systemic chemotherapy, surgery being only a catch-up
74 solution in case of persistent tumour remnants at the end of treatment.

75

76 **Keywords : Myeloid sarcoma, Chloroma, Acute Myeloid Leukaemia ;**
77 **Extramedullary ; Oral tumors.**

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80 **Conflict of Interest Statement: None**

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114 **Captions to illustrations**

115

116 **Figure 1 :** Clinical view of the palatine swelling

117

118 **Figure 2 :** Coronal T1 with gadolinium MRI acquisition showing the palatine mass of 32mm
119 diameter without mass effect.



