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CASE REPORT

Unilateral parotid gland swelling as the sole presenting symptom of acute lymphoblastic leukaemia in children

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KEYWORDS

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Abstract Acute lymphoblastic leukaemia (ALL) in infants below 1 year of age contributed 2.5–5% of childhood ALL incidence. Children with ALL commonly presented with fever, bruising, mucosal bleeding, bone pain, pallor, hepatosplenomegaly, and lymphadenopathy. Common extra medullary leukaemic infiltration has also been reported at diagnosis of ALL to sites such as the liver, spleen, lymph nodes, brain, testes and even nephromegaly. However ALL presented with parotid infiltration is exceedingly rare. We herein present a case of unilateral parotid enlargement in a child with newly diagnosed ALL. This unusual presentation focuses on the importance of considering ALL in the differential diagnosis of parotid enlargement especially when associated with abnormal blood counts.

1. Introduction

Parotid gland enlargement in children is most commonly secondary to infectious and inflammatory lesions.[1] Acute lymphoblastic leukaemia (ALL) is the most common malignancy in children and accounts for 25% of all childhood cancers.[2] Leukaemia typically presents with non-specific symptoms and signs such as anorexia, fatigue and irritability. As the disease progresses, other signs and symptoms such as pallor, bleeding tendency, hepatosplenomegaly and lymphadenopathy may appear. Uncommonly, joint pain, proptosis, abdominal pain, malena, diarrhoea, dysphagia can also be noted as initial manifestations, which may bewilder the clinician as well as the pathologists.[3] Leukaemia may cause enlargement in the salivary glands with or without lacrimal gland involvement.

Primary neoplasms of the parotid gland are rarely seen in childhood. Parotid swelling seen in ALL could be due to

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infiltration of blasts into the parotid gland, which may mimic mumps.[4] Parotid gland involvement in acute myeloid leukaemia (AML) has been reported previously as an uncommon presenting manifestation or relapse of disease. However parotid gland enlargement as a presenting manifestation of acute lymphoblastic leukaemia (ALL) is very rare.[5]

2. Case summary

A 2-year-old boy presented to Otorhinolaryngology (ORL) Clinic with a complaint of painless right parotid swelling for 1 week duration with no history of toothache or trauma. No pallor, fever or other symptoms were documented. Examination revealed right parotid mass, size 2 × 1 cm which was firm in consistency, non-tender and with no surrounding skin changes. Facial nerve examination showed no abnormality. Oral cavity, oropharynx and otoscopic examination showed normal findings. Full blood counts showed haemoglobin 12.3 g/L, TWBC 14.2/L and platelets 459/L. Erythrocyte sedimentation rate is 10 mm/h. An ultrasonography of the neck and parotid region showed an irregular multiseptated cystic intra parotid lesion size 2.4 cm × 2.0 cm × 1.6 cm. Upon Doppler study no colour flow was seen. Multiple enlarged bilateral cervical lymphadenopathy with variable sizes are also seen in ultrasound of neck and parotid.

Further investigation with Magnetic Resonance Imaging (MRI) of head and neck showed enlargement of the right parotid gland. There is a well-defined and homogenous lesion measuring 2.0 cm × 4.4 cm × 3.8 cm involving the superficial and the deep lobe of the right parotid gland, and it envelopes the mandibular vessels. There is also an intraluminal thrombus measuring 0.4 cm in diameter and 2.0 cm in length within the right internal jugular vein with bilateral cervical lymphadenitis. The lesion in MRI was most likely representing parotitis. Subsequent fine needle aspiration cytology (FNAC) of the right parotid mass reported atypical cells seen suspicious of lymphoma. Following this, 2 weeks later, the child underwent right superficial parotidectomy and post-operative recovery was good with no facial nerve palsy. Histopathologic analysis from the right parotid mass specimen confirmed its diagnosis as acute lymphoblastic leukaemia (Fig. 1).

Immunohistochemical stains done to the neoplastic cells showed positive for CD79 alpha, strongly positive for Pax 5, CD10, CD34 and Tdt, (Fig. 2). They are negative for CD20, CD3, CD5, CD117 and LCA. The proliferative index marker (Ki67) is 50–60%. Upon confirmation of the diagnosis, patient was then referred to paediatric haematology team. A bone marrow trephine finding which was performed later revealed thatmarrow cells are consistent with acute lymphoblastic leukaemia.

3. Discussion

In the paediatric population parotid neoplasms are very rare, with benign tumours accounting for 80% of the masses in the parotid gland. Incidence of malignant parotid tumours in the paediatric population is even rarer. Mucoepidermoid carcinoma is the most common malignant neoplasm found in paediatric patients. The parotid gland has been described as a site of extramedullary relapse in patients with primary ALL and AML. It is also seen in newly diagnosed AML in children and adults with newly diagnosed T-cell leukaemia and newly diagnosed lymphoma.[1] Some literature also described the incidence of secondary neoplasms of the parotid gland (including mucoepidermoid carcinoma, acinar cell carcinoma) in patients previously treated for childhood ALL and AML.[1,2] However, not much is known about the involvement of the parotid gland in newly diagnosed ALL.[1]

There are reports on relapse of disease or secondary malignancy in the parotid/salivary glands after treatment of ALL or AML but ALL presenting with parotid mass, although it has been mentioned previously is infrequent, and lacked supportive radiologic evidence.[1,3,6] Parotid gland has been reported to be a sanctuary site for the relapse of AML either in isolation, or in association with the central nervous system but the significance of parotid infiltration by lymphoblasts is uncertain.[5] Parotid gland involvement in ALL patients at initial presentation is possibly a part of bulky extramedullary disease. It is

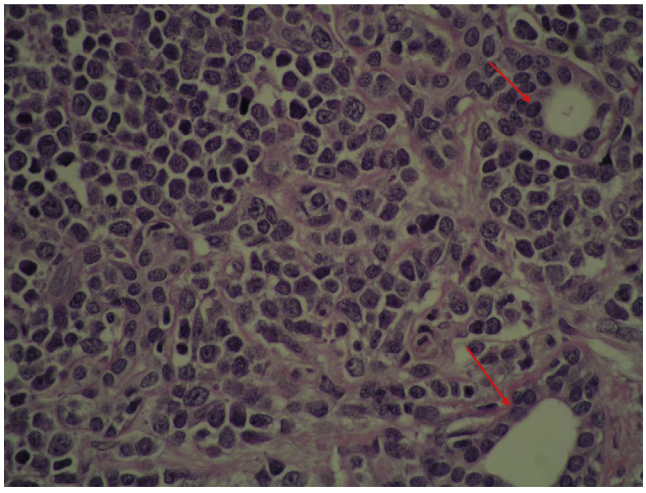

Figure 1a Diffuse atypical lymphoid cells with occasional residual salivary ducts (arrows).

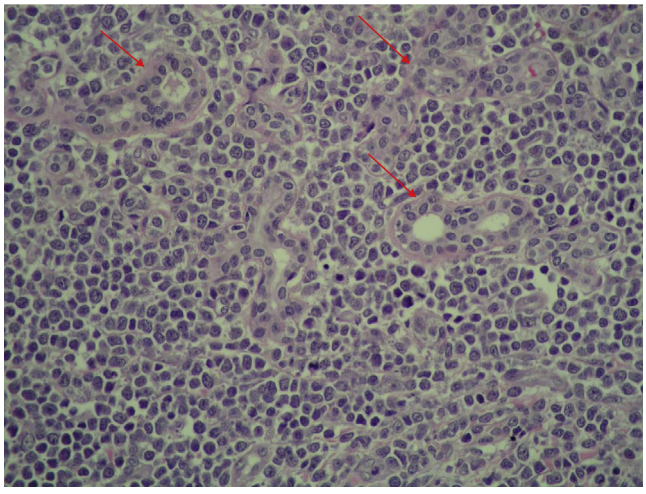

Figure 1b Diffuse atypical lymphoid cells consisting of small to medium cells with irregular nuclei and a few residual salivary ducts (arrows).