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CASE REPORT

An adult juvenile xanthogranuloma in the buccal mucosa

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Abstract Juvenile xanthogranulomas (JXGs) are a type of non-Langerhans cell histiocytosis that commonly affects infants and children. Adult oral JXGs are very rare. A 32-year-old Taiwanese male presented with the chief complaint of a solitary, firm, painless, non-tender swelling over the right buccal mucosa for about 2 weeks. An excisional biopsy of the lesion revealed a mixture of histiocytes, inflammatory cells, and Touton giant cells, and immunohistochemical positivity for CD68 and negativity for S-100 and CD1a confirmed the diagnosis of a JXG. Therefore, the current case report documents, to our knowledge, the first occurrence of an adult oral JXG in the buccal mucosa. It is also the first case of an adult oral JXG to be reported from Taiwan. The clinical characteristics of adult oral JXGs are also briefly reviewed.

Introduction

According to the Histiocyte Society, histiocytic conditions are categorized into Class 1, Langerhans cell histiocytoses (LCHs); Class 2, non-LCHs; and Class 3, malignant histiocytoses.[1] Among these three categories, Class 2 consists of non-LCH disorders of childhood, of which juvenile xanthogranulomas (JXGs) are the most frequent disorder[2]; it

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also encompasses indeterminate cell histiocytoses and sinus histiocytosis with massive lymphadenopathy (Rosai-Dorfman disease), papular xanthomas, progressive nodular histiocytomas, generalized eruptive histiocytomas, and benign cephalic histiocytosis.[1]

The most common locations of JXGs are the skin, predominantly the scalp and face, followed by the trunk, upper extremities, and lower extremities.[3] Visceral lesions, despite being rare, have also been reported, with the predominant regions affected being the eyes and testes.[3] Oral mucosal lesions are uncommon, and only about 30 histologically proven cases have previously been reported in the English-language literature.[4] JXGs usually occur in infants and children, frequently in those younger than 1 year of age.[4] Furthermore, a bimodal climax was reported in those either younger than 1 year or in the fourth decade of life.[4] In a review of the English literature, reports of oral adult JXGs are very rare, there having been only seven previously reported cases.[4–10] The objective of the present article is to describe, to our knowledge, the first case of adult oral JXG occurring in the buccal mucosa. In addition, this article describes the first case of an adult oral JXG reported from Taiwan. The clinical features of adult oral JXGs are briefly reviewed.

Case report

A 32-year-old Taiwanese male visited our institution complaining of a painless, non-tender, smooth-surfaced, firm, pinkish swelling, measuring about 2 × 2 cm in diameter, in the right buccal mucosa for 2 weeks. The overlying mucosa was intact and fixed to the underlying connective tissue. The patient had no history of trauma or infection, and his medical history was non-contributory. All routine laboratory results were within normal limits, and no other skin or visceral lesions were present. Regional lymph nodes were non-palpable. Differential clinical diagnoses included fibrous hyperplasia and a minor salivary gland tumor. An excisional biopsy was performed under local anesthesia, and the tissue was subsequently histologically examined. The biopsy site healed uneventfully, and the patient has been followed-up for 1 year, with neither recurrence nor additional lesions at cutaneous sites.

A gross examination showed a roundish, non-encapsulated soft-tissue specimen of about 1.5 cm in diameter. The cut surface of the lesion revealed a homogeneous whitish appearance. Histologically, a mixture of histiocytes, inflammatory cells, and Touton giant cells with a wreath-like nuclear arrangement of histiocytes was noted (Fig. 1A and B). Specific stains for acid-fast bacilli, fungi, and mucin were negative, while immunohistochemical (IHC) staining was positive for CD68 (Fig. 2A) in tumor cells but negative for CD1a and S-100 (Fig. 2B). No mitotic activity, necrosis, epidermal involvement, fibrous proliferation, significant cellular pleomorphism, or nuclear atypia were observed. These microscopic findings were consistent with a JXG in the buccal mucosa in an adult.

Discussion

JXG was first reported in 1905 by Adamson, who defined single or multiple cutaneous nodules of infancy as a congenital xanthoma multiplex.[11] The term JXG was proposed in 1954 by Helwig and Hackney,[12] and despite the occurrence of adult cases of JXG, this nomenclature is still used today.

The first case of a cutaneous adult JXG was described in 1963 by Gartmann and Tritsch,[13] and about 10–30% of cases of JXG occur in adults,[14] with the peak incidence being in the late 20s to early 30s, with an even sex distribution.[9] The first adult oral case was found in the palatal gingiva by Takeda et al in 1986[5]; since then, only eight oral cases (including the present case) of adult JXG have been reported.[4–10]

The clinical characteristics of the eight reported adult oral JXGs are summarized in Table 1.[4–10] The mean age of occurrence in these oral adult cases was 47.9 (range, 32–64 years); three patients were over 60 years of age.[7–9] Two of the adult oral cases occurred in Asia (one patient was Japanese,[5] the other, the case described in this report, was Taiwanese); another two occurred in whites[4,10]; while four patients were of unknown race.[6–9] In the reported cases, two JGXs were located in the gingiva[4,5] and two in the tongue[6,7]; one occurred in the upper lip[9]; and the remaining three cases occurred in the intra-masseteric muscle,[10] mandibular alveolar mucosa,[8] and buccal mucosa (present case). Therefore, the current case, to our knowledge, is the

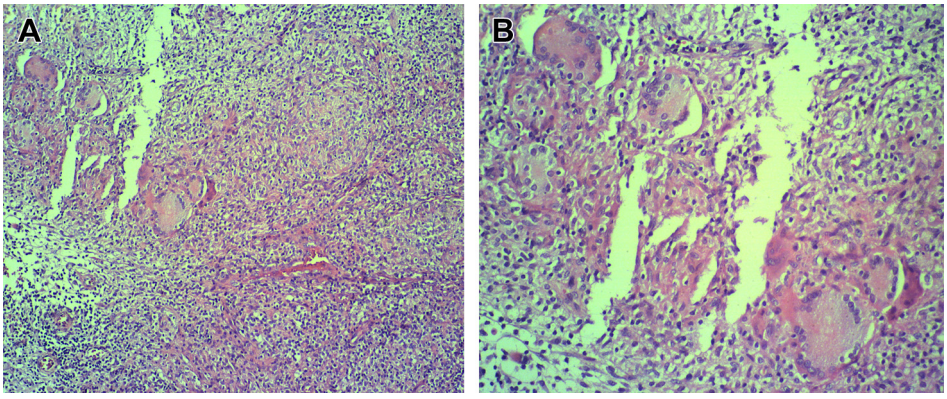

Figure 1 Hematoxylin and eosin staining revealed characteristic microscopic features of a juvenile xanthogranuloma and the presence of histiocytes and Touton giant cells: (A) 100×; (B) 200×magnification.