AN UNUSUAL PRESENTATION OF ADENOID CYSTIC CARCINOMA

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ABSTRACT

Adenoid cystic carcinoma (ACC) is a locally aggressive salivary gland neoplasm, which has a poor long-term prognosis. ACCs are characterized by locally aggressive nature, high recurrence rate, perineural invasion and distant metastases, especially to the lungs and bones. Most common site of involvement is palate and it will present as swelling or growth. Other than conventional radiographs MRI, USG and CT are advised to assess extent and metastasis of the tumour. Although primary treatment is surgical removal, adjuvant radiotherapy is also given following surgery.

Keywords: Adenoid Cystic Carcinoma; Lip; Salivary Gland; Tumour

Introduction

Adenoid cystic carcinoma (ACC), first described as “cylindroma” by Billroth in 1859,1 is a malignant tumour that is commonly classified with the salivary gland tumours although it may arise in any site where secretory glands exist. Overall, adenoid cystic carcinoma is a rare tumour, accounting for only 1% of all malignant tumours of the oral and maxillofacial region and 20-25% of malignant salivary gland tumours.2,3 Fifty per cent of all ACCs occur introraally, frequently seen in the hard palate region.4,5 Herein we report a case distinguished by its uncharacteristic clinical presentation in a 46 year old male patient. Patient presented with a Non-healing ulcer on lip for last two years. This case was different from the cases reported in the literature because of the uncommon site as well as presentation.

Case report

A 46 year old male patient, farmer by occupation, reported to the Department of Oral Medicine and Radiology with a chief complaint of non-healing ulcer on the right side of the upper lip for last two years. Patient noticed a small nodular swelling on the right side of the upper lip two years back. He manipulated the swelling that subsequently led to the formation of an ulcer. The ulcer was small in size to begin with and increased in size over a period of time. Ulcer was tender with indurated margins and base medially to the retro commissural area (Figure 1). The edge of lip revealed an ulcer measuring about 2cm × 1.5 cm on the right side of the upper lip, extending laterally to skin and medially to the retro commissural area. The ulcer was tender and firm in consistency. On examination of lip revealed an ulcer measuring about 2cm × 1.5 cm on the right side of the upper lip, extending laterally to skin and medially to the retro commissural area (Figure 1). The edge of lip revealed an ulcer measuring about 2cm × 1.5 cm on the right side of the upper lip, extending laterally to skin and medially to the retro commissural area (Figure 1). The ulcer was tender and firm in consistency. On examination, the ulcer was tender with indurated margins and base and it was not fixed to the underlying structures. There were no signs of anesthesia or paraesthesia surrounding the ulcer.

Extra oral examination revealed a single, hard, fixed and non tender lymph node palpable in the right submandibular region. Three lymph nodes were palpable in sub mental region among which one was hard and fixed. The rest two were joined together, tender and firm in consistency. On examination of lip revealed an ulcer measuring about 2cm × 1.5 cm on the right side of the upper lip, extending laterally to skin and medially to the retro commissural area (Figure 1). The edge of lip revealed an ulcer measuring about 2cm × 1.5 cm on the right side of the upper lip, extending laterally to skin and medially to the retro commissural area (Figure 1). The ulcer was tender and firm in consistency. On examination, the ulcer was tender with indurated margins and base and it was not fixed to the underlying structures. There were no signs of anesthesia or paraesthesia surrounding the ulcer.

Based on the history and clinical examination, provisional diagnosis of carcinoma of lip was considered. Clinical differential diagnosis included tuberculosis ulcer and basal cell carcinoma. Later patient was subjected to investigations including chest X ray, FNAC of lymph node and incisional biopsy. Chest X ray revealed no significant radiographic changes (Figure 2) and the FNAC smear of submandibular lymph node showed lymphocytes and few histiocytes suggestive of reactive hyperplasia.

Histopathology shows proliferation of monomorphic darkly stained cells in the form of tubular and trabecular pattern with few scattered duct like pattern. Basal lamina like substrate surrounding the tumor islands was suggestive of salivary gland origin (Figure 3). Histopathological features were consistent with Adenoid cystic carcinoma. Treatment plan included surgical excision of the tumour under general anesthesia with follow up.

Discussion

Minor salivary gland tumors occur less frequently than major salivary gland tumors and upper lip is a relatively uncommon site for salivary gland neoplasm.6 However, those originating from the minor salivary glands are mostly malignant tumors.7 As the size of the salivary gland decreases, chances for malignancy increase. Adenoid cystic carcinoma most often presents a diagnostic and treatment challenge owing to the rarity of the lesion. The WHO definition of ACC is, “A basaloid tumor consisting of epithelial and myoepithelial cells in various morphological configurations including tubular, cribriform and solid patterns. It has a relentless clinical course and, usually, a fatal outcome.”8,9

Incidence of ACC is highest in the fifth and sixth decades of life and there is slight female sex predilection. In our case the patient was male and he was in fifth decade of life. Palate is the common site for ACC arising from minor salivary glands and intra oral tumours may possibly show mucosal ulceration. The clinical presentation of ACC involves a slow growing, firm, uni-lobular mass.8,9 Pain is usually a commonly associated symptom, may occur prior to the clinical evidence of the disease. Our case was presented as a non healing ulcer on the upper lip, which is not a common site and usual presentation seen in ACC as reported by literature.11 Such ulcers are often mis-
An unusual presentation of Adenoid Cystic Carcinoma

Histologically, ACC has three variants - cribriform, tubular and solid. ACCs are graded according to the histological pattern into grade I, grade II and grade III with Grade I being a combination of cribriform and tubular, Grade II a mixture of cribriform, tubular and solid patterns and Grade III having only solid pattern. Prognosis of ACC is influenced by many factors. These include staging of tumor, positive surgical margins, and site of involvement, perineural invasion, histological type and presence of cervical lymph node metastasis at the time of diagnosis.

Conclusion
In conclusion, a careful history, examination and proper investigations are essential for the successful management.

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References

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