

CASE REPORT

Intraosseous sebaceous adenocarcinoma of the maxilla

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Abstract

Primary intraosseous salivary malignancies are rare. The adenoid cystic adenocarcinoma, adenocarcinoma not otherwise specified, acinic cell adenocarcinoma and the epithelial-myoeplithelial carcinoma have all been reported within the jaws. The mucoepidermoid carcinoma is by far the most common tumor in this setting accounting for 2%-4% of all mucoepidermoid carcinomas. An intraosseous sebaceous adenocarcinoma has never been reported. We document a case of sebaceous adenocarcinoma of the maxilla in a 56-year-old woman who presented with a painful swelling of the right face. CT scans showed an expansile, osteolytic tumor of the right maxilla. An exploratory biopsy revealed the tumor to be expanding and eroding the cortical plates including the floor of the sinus. The histopathology was consistent with a sebaceous adenocarcinoma. The sebocytes expressed EMA and CD15. The basaloid cells were CD15 negative. The tumor was treated with a wide local resection. Post-surgical healing was uneventful and patient is being monitored regularly for recurrences. Prognosis of any tumor originating intraosseously is worse than its soft tissue counterpart. Extraocular sebaceous adenocarcinomas are low grade malignancies which can recur locally. As this is the first report of an intraosseous/central sebaceous adenocarcinoma, prognostication could be inaccurate and therefore long-term follow-up is highly recommended.

Key words

Intraosseous tumor, Sebaceous adenocarcinoma, Maxilla

1 Introduction

Primary intraosseous malignancies of glandular differentiation are rare. They constitute 0.4% of all salivary gland malignancies. They have wide histologic variability and the etiology is poorly understood. They present with swelling, pain and paresthesia. The mucoepidermoid carcinoma, adenoid cystic adenocarcinoma, adenocarcinoma NOS, acinic cell adenocarcinoma and the epithelial-myoeplithelial carcinoma have all been reported within the jaws^[1,2]. We document an interesting case of sebaceous adenocarcinoma arising in the maxilla. This malignancy arising in an intrabony location has never been reported.

2 Case presentation

A 56-year-old woman presented with a painful swelling on the right side of her face. CT scans showed an expansile, osteolytic tumor of the right maxilla (see Figure 1). An exploratory biopsy revealed the tumor to be expanding and eroding the cortical plates including the floor of the sinus but with no extension into the sinus epithelium (see Figure 2). Histopathological examination of the mass showed islands of neoplastic cells with a basaloid cytomorphology at the periphery and cells with clear or multivesicular cytoplasm evolving towards the center suggestive of sebocytic differentiation (see Figure 3). This biphasic pattern was highly suggestive of a sebaceous adenocarcinoma. The ratio of basaloid cells to sebocytes was highly variable and some sheets of tumor showed a preponderance of basaloid cells (see Figure 4). Rarely, a peripheral palisading was also noted (see Figure 5). Some islands also showed comedo-type necrosis (see Figure 6).

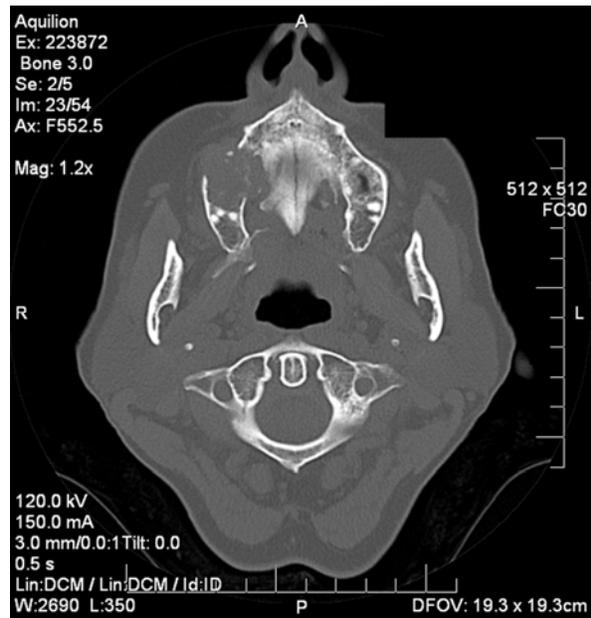


Figure 1. CT image (axial view) showing an expansile, osteolytic lesion arising in the right maxilla

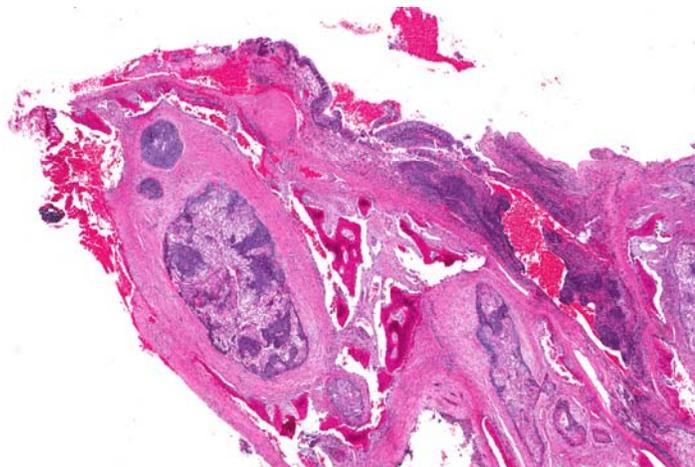


Figure 2. Tumor islands eroding the cortical bone of the floor of the sinus (H&E stain. 20×)

Immunohistochemically, EMA highlighted areas of sebocytic differentiation. Many sebocytes exhibited accentuated EMA expression overlying the cytoplasmic vesiculation (see Figure 7). CD15 was also used to highlight the sebocytes. The basaloid cells did not stain with CD15 (see Figure 8). The tumor was removed with a wide local resection including a hemimaxillectomy. Post-surgical healing was uneventful and the patient is being monitored regularly for recurrences.

Figure 3. Nests of varying size consisting of dark basaloid cells and vacuolated to microvesicular cytoplasm rich sebocytes (H&E stain. 40×)

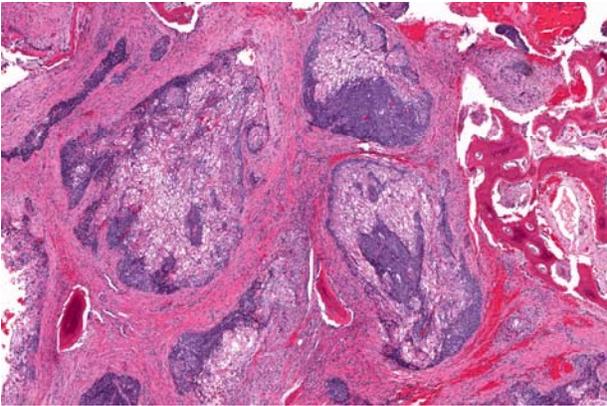


Figure 4. Some islands were predominantly basaloid with scattered squamoid, vacuolated cells or cells with a microvesicular cytoplasm (sebocytes) (H&E. 200×)

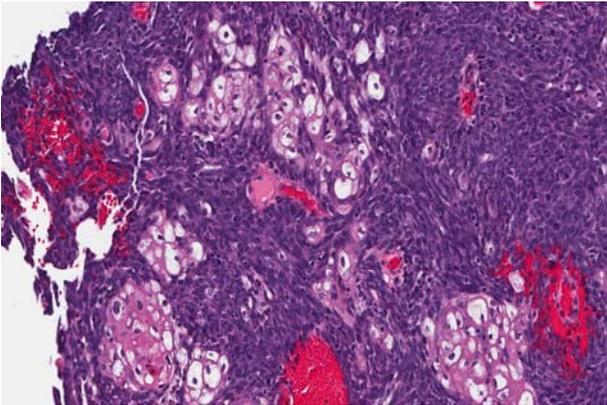


Figure 5. Tumor cell nest demonstrating some peripheral palisading (H&E. 100×)

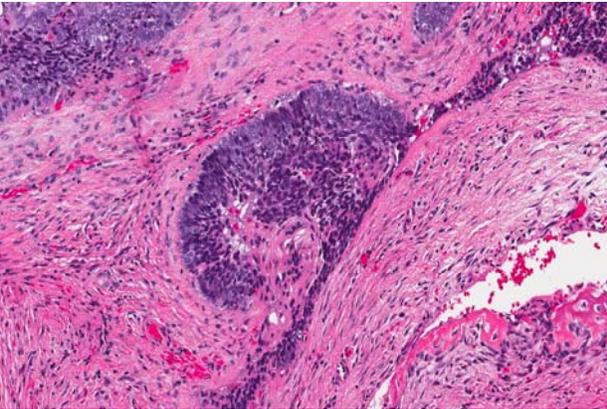
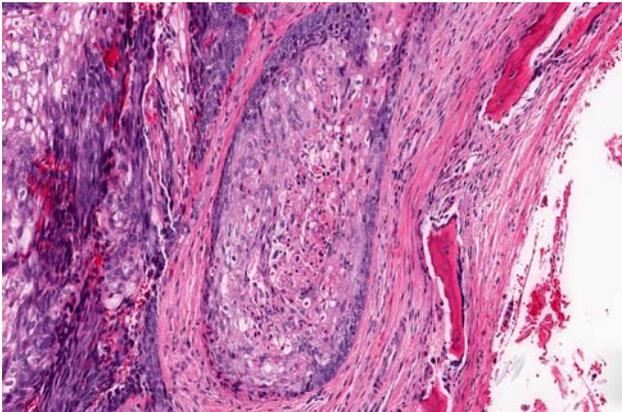


Figure 6. Tumor island showing comedo-type necrosis (H&E. 100×)



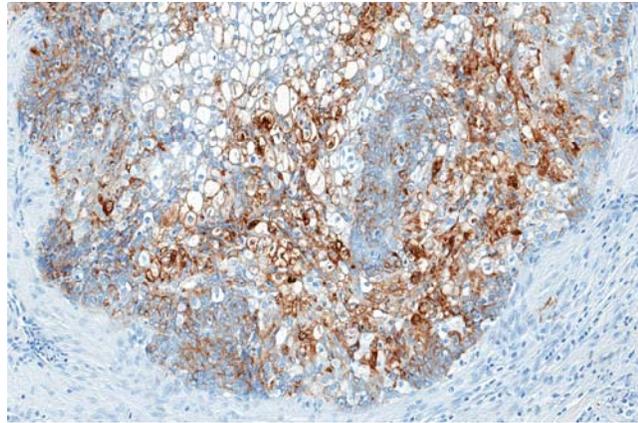


Figure 7. EMA highlights the sebocytes with some sebocytes also showing cytoplasmic accentuation

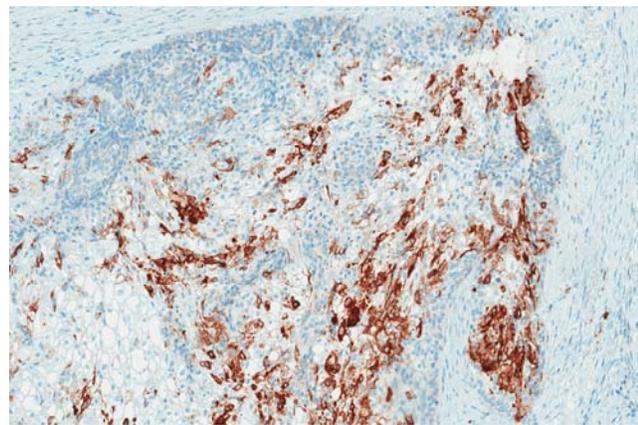


Figure 8. Sebocytes express CD15 reactivity while basaloid cells are non-reactive

3 Discussion

Primary intraosseous salivary malignancies are uncommon. They are known to occur primarily in the mandible and very few arise in the maxilla. Mucoepidermoid carcinoma is the most common intraosseous (central) malignancy of salivary gland differentiation accounting for 2%-4% of all mucoepidermoid carcinomas^[1,2]. There are no reports of the occurrence of the sebaceous adenocarcinoma in the maxilla.

All the criteria for a primary intraosseous (central) salivary gland neoplasm such as radiologic evidence of osteolysis, expansion of cortical plates, intact mucous membranes, absence of any salivary glands and histologic evidence of the morphology of the tumor were fulfilled in this case^[3]. While the pathogenesis is uncertain, several theories have been proposed. Entrapment of the salivary glands, mucous glands and sebaceous glands during embryonic development of the jaw, neoplastic transformation of odontogenic epithelium or sinus epithelium to list a few^[4].

Brookstone and Huvos established a staging system for central salivary gland malignancies based on bony invasion – Stage 1: Lesions with intact cortical plates with no evident bony expansion; Stage 2: Tumors with intact plates, but intrabony expansion; Stage 3: Lesions associated with cortical perforation or nodal disease^[4]. As per these criteria, the present case was classified as stage 3.

Sebaceous glands are known to occur in ectopic locations (Fordyce’s granules) but rarely become neoplastic. Sebaceous carcinoma occurs most frequently in periocular area including the eyelids and these tumors are unencapsulated and characterized by infiltrating nests of cells with varying degree of sebaceous differentiation, marked nuclear atypia, abundant mitosis and a locally infiltrative growth pattern. The tumor cells form sheets, irregular nests, trabeculae and cords invading into the surrounding tissue and around nerves. Large tumor nests may also show comedo necrosis.

Sebocytic differentiation is highly variable from small clusters to large islands. In most cases, the sebocytes are in the center of islands, nests or sheets of epithelial cells. The epithelial cells predominate and may be basaloid or squamous. The sheets, islands and nests may show a vague peripheral palisading. Isolated mucocytes may also be noted among the tumor cells. Some sheets and islands of tumor cells may also demonstrate cystic areas.

Immunohistochemically, sebocytic differentiation is highlighted by their reactivity to several epithelial markers including EMA, BerEP4, cytokeratin 7, CAM 5.2 and BRST-1. EMA accentuates the cytoplasmic vesiculation of sebocytes as shown in our Figure 7. Sebocytes also react with CD15, androgen receptor and gross cystic disease fluid protein 15 (BRST-2). It is not established if reactivity to androgen receptor protein is helpful in excluding other clear-cell tumors. The basaloid or squamoid epithelial cells do not stain with these markers. Sebocytes are non-reactive to mesenchymal, neural, and melanocytic markers. In frozen sections, the sebocytes express Oil Red O and Sudan black ^[5].

Molecular studies show that mutations in TP53 gene is seen in invasive sebaceous carcinoma and not in situ carcinoma. Certain strains of human papilloma virus have been associated with the pathogenesis of sebaceous carcinoma ^[6]. Rarely, sebaceous neoplasms may be associated with Muir-Torre syndrome. Muir-Torre syndrome is a rare autosomal dominant disease that is characterized by the development of multiple benign and malignant cutaneous sebaceous neoplasms, keratoacanthomas, and visceral cancers, usually of gastrointestinal origin. The sebaceous tumors are frequently the initial manifestation of the syndrome ^[7].

Wide surgical excision with or without chemo/radiation therapy has been the treatment of choice in extraocular locations and elective neck dissection is performed for tumors with a poor prognosis ^[7].

Sebaceous carcinoma is considered an intermediate grade malignancy with 5-year survival rates from 60%-70% with a 30% rate of recurrence ^[7]. It is difficult to grade or prognosticate the central intraosseous tumors in the same way as salivary gland or eyelid tumors as the prognosis of any tumor arising ectopic in the bone is typically less favorable. Extraocular sebaceous adenocarcinoma are low grade malignancies which can recur locally. As this is the first report of an intraosseous/central sebaceous adenocarcinoma, prognostication could be inaccurate and therefore upon removal long term follow up is highly recommended.

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