Sarcomatoid Carcinoma of the Hard Palate: An Unusual Cause of Palatal Ulcer

Sir,

Sarcomatoid (spindle cell) carcinoma of the head and neck, a subtype of squamous cell carcinoma, is a rare biphasic neoplasm. It has more aggressive behaviour as compared to classical squamous cell carcinoma and rarely presents as a palatal ulcer. We report the case of a 23-year-old female who presented with ulceroproliferative growth on the hard palate, gum hyperplasia, blackening of teeth, facial swelling, and pain.

The patient initially noticed a painful crusted erythematous papule on the anterior aspect of the hard palate 1.5 years back. The lesion progressively increased in size and was associated with diffuse swelling of central face and persistent pain in the head and right paranasal area. She also had epiphora from both eyes. However, there was no history of bleeding, foul discharge or epistaxis. She had mild anorexia but did not complain of weight loss and fever. There was no history of tobacco chewing, smoking, alcohol use, previous irradiation, or poor oral hygiene. There was no history of hematuria, breathing difficulty, or palpable purpura. On examination there was an ill-defined polypoidal, moderately tender, soft-to-firm 4 × 4 cm ulceroproliferative growth on the hard palate covered with white adherent slough [Figure 1]. There were softer, non-friable polypoidal growths involving the upper gingiva along with destruction of alveolar margins. In addition, there was a polypoidal growth on the right lateral nasal wall. There was no significant lymphadenopathy or organomegaly. We kept differentials of Wegener’s granulomatosis, malignancy, tubercular ulcer, rhinosporidiosis, and deep fungal infections. Her laboratory evaluation revealed anemia (hemoglobin: 9.6). Routine urine examination revealed mild hematuria (2–3 red blood cells per high power field). Cytoplasmic antineutrophil cytoplasmic antibodies was weakly positive (1:20). Staining for acid fast bacilli, KOH, and Gram’s stain; gene expert; and culture for tuberculosis was negative. Her mucosal biopsy revealed focally ulcerated hyperplastic stratified squamous epithelium [Figure 2a], with dense surface acute inflammatory exudate, fibrin deposition and large areas of necrosis. Subepithelium showed spindle cell proliferation [Figure 2b and c], with nuclear pleomorphism and increased mitotic activity [Figure 2d]. Tumor cells were positive for cytokeratin and S100, and negative for human melanoma black 45 and smooth muscle actin [Figure 3]. Overall features were those of sarcomatoid carcinoma. Contrast-enhanced computed tomography of the head revealed ill-defined heterogeneous soft tissue thickening with geographic lytic destruction of both the palatal processes of maxilla and alveolar ridges overlying maxillary sinuses [Figure 4]. Positron emission tomography scan showed metabolically active primary pathology in the hard palate with metastasis to left cervical lymph node. The patient was transferred to surgical oncology for further management where she was advised surgery followed by radiotherapy.

Discussion

Squamous cell carcinoma has many morphological and
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histopathological variants, which include verrucous, spindle-cell (sarcomatoid), basailoid exophytic or papillary, clear cell, pigmented, follicular, and adenosquamous carcinoma. Management approach varies with different histopathological variants.[1] Of the many hypotheses for the pathogenesis of sarcomatoid carcinoma, 3 dominant pathogenetic theories have emerged, namely, (1) It may be a “collision tumor;” (2) it is a pseudosarcoma, i.e., squamous cell carcinoma with an atypical reactive stroma; and (3) “De-differentiation” to spindle cell morphology.

The mean age at diagnosis of spindle cell carcinoma is 57 years (29–93 years).[2] Most tumors in the head and neck region occur in the larynx. Other sites that may be involved are the lower lip, tongue, and alveolar ridge or gingiva.[3] It presents as exophytic ulceroproliferative polypoid growth with shaggy exudates, however, sessile, nodular, or endophytic configuration has also been described. Radiation, sun exposure, tobacco use, trauma, and alcohol consumption have been implicated as etiological factors in many cases.[3] Histological appearance may vary from case to case and even in the same patient in different areas. The cells may appear like epithelial cells or as atypical mesenchymal cells. The tumor is made of fascicles of anaplastic spindle cells.[3] The treatment of sarcomatoid carcinoma is controversial. It is believed by many that radical dissection along with removal of metastasis is adequate, whereas others believe that postoperative radiotherapy is also mandatory. Immunohistochemical studies of epithelial and mesenchymal markers are also important in the diagnosis of the tumor as well as in differentiating it from true spindle cell sarcoma, malignant myoepithelioma, and melanoma.

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Conflicts of interest
There are no conflicts of interest.

Sweta Subhadarshani, Savita Yadav, Ashok Singh¹, Anurag Chahal²
Departments of Dermatology Venereology and Leprology, ¹Pathology and ²Radiology, AIIMS, New Delhi, India
A Sporadic Case of Ichthyosis Hystrix: Curth and Macklin Type

Sir,

A 13-year-old female presented with generalized skin lesions since early childhood. She was born healthy out of non-consanguineous marriage and full-term vaginal delivery, according to history given by her mother. After one year, she started developing lesions initially over the scalp with mild scaling. Patients may present at birth or there may be a delayed onset in infancy of non-consanguineous marriage and full-term vaginal delivery, according to history given by her mother. After 1 year, she started developing lesions initially over the scalp with mild scaling. Patients may present at birth or there may be a delayed onset in infancy. The distribution of lesions, associated abnormalities, and inheritance patterns help to distinguish between congenital ichthyosis, and is also known as systematized palmoplantar keratoderma leading to deep, bleeding, and painful fissures. Electron microscopy shows perinuclear vacuolization and the form of crest-like projections of epidermis. Ichthyosis hystrix of Curth and Macklin type is a rare autosomal dominant disorder with very few sporadic cases documented between 1731 and 1851 in the Lambert family of Suffolk. The word hystrix means porcupine-like. It was first described by Brocq in 1875;101:63-6.

References