An Uncommon Malignant Neoplasm of the Caruncle

Report of a Case of Undifferentiated Carcinoma

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The caruncle is an uncommon site for the occurrence of neoplasm despite its diverse histological composition of conjunctival, lacrimal, and skin tissues. The following case report describes a 66-year-old man who suffered from a primary undifferentiated carcinoma of the left caruncle. He remained well with no evidence of recurrence 24 months after a complete surgical excision. To our knowledge, this could represent the first case of undifferentiated carcinoma of the caruncle to be reported in the English literature. Early detection and complete excision for this type of lesion could lead to a satisfactory clinical outcome.

Lesions involving the caruncle are rare; however, a malignant neoplasm of the caruncle is even rarer. Luthra et al reported only 112 caruncular lesions, 4 of which were malignant neoplasm, in a clinico-pathological review during a 52-year period. Other series have similarly documented an overall 4% frequency of malignant neoplasm among caruncular lesions. The more commonly occurring caruncular tumors are nevus and papilloma, while sebaceous carcinoma is the most common malignant tumor. Other rare tumors include malignant melanoma, oncocytoma, mucopidermoid carcinoma, sebaceous carcinoma, adenocarcinoma, and tumors of mesenchymal or vascular origin. We report a rare case of primary undifferentiated carcinoma of the caruncle.

REPORT OF A CASE

A 66-year-old man had a 3-month history of progressive enlargement of a reddish nodular lesion over the left caruncular region (Figure 1). The firm erythematous mass was well circumscribed, measuring about 2.5×1.5×2.0 cm. It was not adherent to the orbital bone or the deeper structures. The upper and the lower canaliculi, including their punctal openings, were uninvolved. No surrounding satellite lesions were found. Visual acuity was 20/50 OD and 10/200 OS. There were bilateral cataracts, with the left cataract being more severe. The patient also had old trachoma with some corneal scar in the left eye. No other ocular abnormalities or regional adenopathy were found. Medical history and systemic physical examination findings were unrevealing. A computed tomographic scan of the orbit revealed a large tumor in the anteromedial aspect of the orbit indenting and displacing the globe to the left (Figure 2). There was no bony involvement and the adjacent nasal cavities were clear. Results of systemic investigations for disseminated malignant neoplasms were normal. The serum IgA titer for Epstein-Barr virus viral capsid antigen was negative. A complete excision of the tumor was performed under general anesthesia. The surgical margins, assessed by frozen section were declared to be free of tumors. No adjuvant chemotherapy or radiotherapy was given. The patient was symptom free and there was no evidence of recurrence or metastasis at the latest follow-up, 24 months after the operation.

Histological examination showed an infiltrative tumor comprising clusters and
lobules of undifferentiated carcinoma cells lying beneath the stratified squamous epithelium (Figure 3). The tumor cells consisted of marked nuclear pleomorphisms, hyperchromasia, distinct nucleoli, and scanty amphophilic cytoplasms (Figure 4). Frequent mitotic activity and apoptosis were evident. No squamous or glandular differentiation was seen and no cytoplasmic vacuolation was identified. Histochemical stain (oil-red-O) for lipid material in frozen section was negative, excluding the possibility of sebaceous carcinoma. Immunohistochemical study findings for epithelial marker (AP1/AE3) were positive while lymphoid marker (leukocyte common antigen), melanocytic marker (S-100 protein), muscle marker (desmin), and neuroendocrine markers (neuron-specific enolase, chromogranin) were all negative. The Epstein-Barr virus–encoded RNAs, which are untranslated RNAs present in Epstein-Barr virus–infected cells, were absent in the specimen.9 The tumor was classified as an undifferentiated carcinoma.

**COMMENT**

The caruncle has a unique embryological origin, with contribution from the lower eyelid and its appendages admixed with the ingrowth from the lower canaliculi.10 When fully developed, the caruncle is lined by non-keratinized epithelium that is similar to conjunctival epithelium, while it harbors all the skin appendageal elements including hair follicles, sebaceous glands, and sweat glands.11 Accessory lacrimal glands can also be seen in this area. As a result, caruncle may spawn any tumor or cyst that occurs in the conjunctiva, skin, or lacrimal gland.12,13 Surprisingly, these are all rare, especially malignant neoplasm, which might be attributed to the small anatomic size of the caruncle.

To our knowledge, this is the first report of undifferentiated carcinoma of the caruncle. We regard this
tumor as a primary carcinoma of caruncle because of the location of the tumor, the exclusion of other primary tumors in the nearby structures by the computed tomographic scan, and the lack of primary tumor found or developed in other parts of the body during the follow-up period. There is, however, at least 1 case each of undifferentiated carcinomas reported in the orbit and maxillary sinus in a 20-year review of orbital exenteration. In addition, 1 case of anaplastic tumor was reported by Pecorella and Garner in their series of caruncular tumors during a 59-year period. No further details or the exact lineage of the tumor was provided. Leung et al reported a case of undifferentiated carcinoma in the lacrimal sac with metastasis to the cervical lymph node at presentation. The tumor was accompanied by intense lymphoid infiltrate and showed the Epstein-Barr virus, compatible with lymphoepithelioma-like carcinoma. The current case does not exhibit the histological features of lymphoepithelioma-like carcinoma and is Epstein-Barr virus negative.

Given the rarity of carcinomas of the caruncle, little is known about their natural history and the plan of management is largely empirical. The mainstay of treatment tends to be surgical excision with orbital exenteration in reserve for late presentation and local recurrence. The efficacy of adjuvant chemotherapy and radiotherapy has not been properly evaluated. In our patient, complete excision with clear surgical margins has achieved a 24-month symptom-free period with no evidence of recurrence. The possibility of late metastasis is still present. Luthra et al have reported a case of sebaceous carcinoma of caruncle that recurred only 10 years after initial excision. Apart from the usual routes of metastatic spread, caruncular tumor may shed along the intracanalicular pathway.

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REFERENCES