Conjunctival myxoma: a case report

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SUMMARY  A rare case of conjunctival myxoma in an 18-year-old female is reported. Clinically it presented as a painless mass located in the nasal bulbar conjunctiva. It was composed of spindle and stellate shaped cells in a loose mucoid stroma. Some of the cells had intracytoplasmic vacuoles consistent with dilated rough endoplasmic reticulum and/or intranuclear vacuoles of nuclear membrane invaginations. Mast cells were also seen in the stroma. No recurrence has been reported eight months postoperatively.

Myxoma is an uncommon benign connective tissue tumour that may arise from various parts of the body.1,2 Myxomas of the ocular adnexa are rare, and only five cases of conjunctival myxoma have been reported.3-7 The following report describes a well-documented case of this rare neoplasm, a conjunctival myxoma, including ultrastructural study.

Case report

An 18-year-old healthy white female complained of a small lesion in the nasal bulbar conjunctiva of her right eye which had been present for four months. There was no history of trauma. The lesion was painless and did not cause any other symptoms, and the patient sought medical consultation only for cosmetic reasons. Examination of both eyes was unremarkable except for a pinkish glistening 'cystic' tumour in the nasal bulbar conjunctiva of the right eye. The tumour did not invade the limbus (Fig. 1). A similar though smaller tumour was found at the same location in the fellow eye. The tumours and the conjunctiva around them moved freely over the sclera. The patient's complaints, however, were limited to the right eye, and under local anaesthesia the tumour was completely removed. After eight months there was no scar or recurrence, and the tumour of the fellow eye has not grown in size.

PATHOLOGICAL FINDINGS

The specimen was composed of a well-circumscribed soft mass measuring 10×4×4 mm. It was composed of a main cyst-like structure, whose cut surface appeared myxomatous, and a connective tissue 'tail.' The entire specimen was covered on one side with a pinkish tissue. A prominent blood vessel, apparently the feeding vessel, was seen entering the tumour (Fig. 2).

Microscopic examination revealed under the slightly acanthotic conjunctival epithelium a mass of very loose connective tissue. It was composed of scattered stellate shaped cells with multiple cytoplasmic processes and spindle shaped cells with bipolar cytoplasmic processes. Both types of cells had moderately large hyperchromatic and slightly pleomorphic nuclei, and were embedded in loose mucoid stroma. The stroma contained reticulin
fibres and sparse small blood vessels, and collagen fibres that were denser in the periphery (Fig. 3). Some of the tumour cells showed tiny nuclear vacuoles, and some had cytoplasmic vacuoles not impinging on the nucleus (Fig. 4). The vacuoles did not stain with oil-red-o, alcian blue, or PAS stains. There was no mitotic activity. A few mast cells were present within the myxoid stroma. The loose stroma contained abundant hyaluronidase-sensitive mucopolysaccharides. Bodian stain was negative for nerve fibres.

Electron microscopic examination disclosed a piece of tumour tissue composed of predominantly fibroblast-like spindle shaped cells that lacked basement membranes, with prominent rough endoplasmic reticulum and very few other cytoplasmic organelles. Many of the nuclei contained one or two
Fig. 5 A cell with infolding of the nuclear membrane, forming membrane bounded vacuoles (v). Note two nucleoli inside the nucleus and prominent rough surfaced endoplasmic reticulum in the cytoplasm (left lower corner) (×12 700). Insert: higher magnification of a vacuole (×34 650).
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Fig. 6 Extremely dilated cisternae of rough surfaced endoplasmic reticulum. Note some collagen fibres outside the cell (×13 500).

nucleoli. The extracellular material was amorphic with sparse collagen fibres. Some cells showed infolding of their nuclear membrane that in some sections formed membrane bounded vacuoles (Fig. 5). In some of the cells the cisterna of the rough surfaced endoplasmic reticulum were markedly dilated (Fig. 6).

Discussion

Pure myxomas have rarely been reported in the ophthalmic literature and only five conjunctival myxomas have been described. Magalif reported the first case of conjunctival myxoma but without any clinical or histopathological description. Mancione described the first case of conjunctival myxoma with clinical and histological details. Fookes described a case that developed close to a filtration scar, and he considered the lesion to be a complication of trephine operation. The later two well-documented cases reported by Doughman and Wenk and Stafford appear to be pure myxomas. The age of patients in the previously reported cases was between 49 and 72 years, while our patient was much younger (18 years). The tumour in all cases was well circumscribed, sometimes described as cystic, and were painless. The size of the tumour in all reported cases, including ours, did not exceed 10 mm in greatest dimension. In two cases the tumour was temporal to the cornea. Follow-up information in two previous cases and in our case showed no recurrence after 9, 64, and 8 months respectively.

The tumour in our case met the criteria of myxoma. It showed relatively small numbers of stellate and spindle shaped cells embedded in a loose stroma with delicate reticulin fibres and sparse vascular structures and delicate collagen fibres. The stroma contained abundant hyaluronidase sensitive mucopolysaccharides. No mitotic activity was present. In addition our case presented some other features that have rarely been described in myxomas—the intracytoplasmic and intranuclear vacuoles. These two features were reported in an intramuscular myxoma. Intranuclear vacuoles alone were described previously in one
case of intramuscular myxoma\(^9\) and in one figure in Stout's study\(^2\) without any textual description, while intracytoplasmic vacuoles alone were described in a case of conjunctival myxoma\(^4\) and a case of intramuscular myxoma.\(^8\) The cytoplasmic vacuoles as seen on electron microscopic examination are apparently consistent with markedly dilated rough endoplasmic reticulum, while the intranuclear vacuoles were found to be invaginations of markedly folded nuclear membrane.

Mast cells, demonstrated by Giemsa stain in our case, have not been described previously.

In the differential diagnosis of pure conjunctival myxoma we have to consider mixed myxomas such as fibromyxoma, lipomyxoma, or fibrolipomyxoma, and myxomatous degeneration of tissue or other tumours such as neurofibroma. The degenerative process is sometimes very difficult to prove. It is more important to consider the possibility of malignant tumours such as myxoid liposarcoma and malignant fibrous histiocytoma.\(^1\)

References