Granular cell tumour vocal cord in an adolescent child - A case Report

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Abstract

Granular cell tumors are benign relatively rare lesions that can occur in any organ in the body. Tongue is the most predilected site. Granular cell tumor vocal cord is even rarer and typically occurs in the third decade of life. It generally presents as a single lesion, but multiple lesions can also occur. Patients present with hoarseness and/or airway obstruction. Granular cell tumors are resistant to radiotherapy, and therefore surgical excision is the treatment of choice. We report the case of granular cell tumor vocal cord in a 13-year-old male child in whom direct laryngoscopic excision of the tumor was advocated.

Keywords Granular cell tumour • Direct laryngoscopy

A 13-year-old boy presented to Department of ENT medical college Kottayam, with history of change in voice and frequent clearing of throat of 5 months duration. He was subjected to detailed ENT evaluation and indirect laryngoscopic examination showed a smooth reddish mass involving the entire length of right vocal cord with normal vocal cord mobility. Neck and other ENT areas were within normal limits. General examination did not reveal any abnormalities. He was investigated and subjected to Direct Laryngoscopy under General Anaesthesia. The laryngoscopic finding was a firm reddish mass involving the entire length of membranous part of right vocal cord which was completely excised and the specimen sent for histopathological examination. Post operative period was uneventful.

The histopathology (Photo 1, 2 and 3) report came as Granular cell tumour. During the first postoperative review clinical examination showed a normal larynx with normal vocal cord and the patient was absolutely free of symptoms. He is under regular follow up now.

Discussion

Granular cell tumour is a benign mesenchymal neoplasm of schwann cell origin. Previously it was named as Granular cell myoblastoma and also called as Abrikossoff tumour. Current etiological data suggest that granular cell tumour is the consequence of degradation and is not a specific neoplastic entity. The granular cell tumour usually affects middle aged individuals of range 30 to 50 years and there is slight female predilection. The oral cavity, specifically the tongue is the most common site of its occurrence. Other sites include the anterior and posterior chest wall, upper limbs, larynx, stomach, vulva and anogenital region. Most common location in larynx is the true vocal cords-middle and posterior 1/3rd.

Clinical features depend on the location and size of the tumour, and that of vocal cord, present as change of voice, dyspnoea and stridor. Tumour may arise simultaneously in other sites also. Histopathologically, it is a non encapsulated tumour

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with poorly defined margins and is composed of sheets, cords, or nest of cells. The cells are large and polygonal with abundant pale eosinophilic cytoplasm and small vesicular nuclei. 50% of cases shows pseudo epitheliomatous hyperplasia (P E H) of the overlying epithelium. PEH is defined as a florid epithelial proliferation with an apparent invasive pattern that mimics infiltrative squamous cell carcinoma, so insufficient deep biopsy may lead to misdiagnosis of well differentiated squamous cell carcinoma.

Immunohistochemistry –The cytoplasmic granules stain positive for periodic acid Schiff and resistant to diastase indicating that the granules are not glycogen. Recent studies suggest they are myelin or myelin break down products. Electron microscopy reveals that the membrane bound cytoplasmic granules to be phagolysosomal complexes. The tumour cell show positivity with S100 protein which is supportive of a neural origin. Neuron specific enolase, Vimentin, PGP95 and melanoma associated antigen (NK/C-3) The pan-macrophage marker CD68 [kp1] reacts strongly with granular cell tumour.

Treatment

The treatment of choice for granular cell tumor is surgical resection. It may be by endoscopic removal or direct laryngoscopic excision. Other methods include Microlaryngoscopic excision using 400 mm objective or Co2 Laser Resection. It is resistant to radiotherapy. Recurrence rate is 2-8%. Follow up during post operative period is important. There is rare chance of malignant transformation. Incidence is 2% of all granular cell tumour. Clinically malignant behavior is suggested by history of local recurrence, and recent rapid growth.

Histologic criteria for assessing the malignant potential are Necrosis, cytologic spindling, vesicular nuclei with large nucleoli, increased mitotic activity [$>2/10$ high power field], high nuclear to cytoplasmic ratio and pleomorphism. If two of these criteria present considered as Atypical and three or more present it is malignant.

Conclusion

Granular cell tumour vocal cord is a rare benign neoplasm with very rare occurrence in children. Only 17 cases reported in literature. Characteristic histopathological feature is pseudo epitheliomatous hyperplasia which mimic well differentiated squamous cell carcinoma. Immuno histochemistry has role in definitive diagnosis. Local Excision can completely cure the disease.

References

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Photo Granular cell tumour vocal cord

- 1: Low power view
- 2: High power view
- 3: Scanner View

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