

Papillary Adenocarcinoma of the Nasopharynx - Case Report and Review of the Literature

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Summary

We report a rare tumour of the nasopharynx- papillary adenocarcinoma. This is usually of low grade and certainly in our patient it behaved so. It is even rarer to have this tumour in a patient with Turner's syndrome in whom there is a high incidence of gynaecological malignancy. It has not previously been documented and the occurrence in this patient is probably coincidental.

Key Words: Papillary adenocarcinoma, Nasopharynx, Turner's syndrome

Introduction

Low grade papillary adenocarcinoma is a rare tumour in the nasopharynx. It has been variously called lobular or terminal duct carcinoma, trabecular adenocarcinoma and polymorphous low grade adenocarcinoma (PLGA). It is thought to arise from the minor salivary glands in the nasopharynx although its exact origin is still being debated and some favour an epithelial origin¹. It's unusual for this to present in the nasopharynx as the large majority occurs in the oral cavity. Reported cases demonstrated 63% involve palate, 14% buccal mucosa, 11% upper lip, 7% retromolar region and 5% other intraoral sites². The differential diagnosis includes seromucous gland tumour of the nasopharynx which is by far the most common tumour at this site. The general consensus on management is the same as that of minor salivary gland at other sites involving surgery followed by radiotherapy for incompletely excised tumour (in the nasopharynx it is generally difficult to obtain adequate margin). Metastatic papillary adenocarcinoma in the nasopharynx is also a very rare entity and has to be excluded. Thyroid primary was

excluded in this case in the light of the negative stains for thyroglobulin and negative thyroid nuclear and ultrasound scans.

Another interesting point is the association with Turner's syndrome in whom there is a propensity to develop gynaecological tumours. We have not come across reported association of low grade papillary adenocarcinoma with Turner's syndrome.

Case report

A 29-year-old Chinese female presented with a 3 months history of left nasal blockage and discharge. There was associated ipsilateral hearing loss and facial weakness. Physical examination showed features of Turner's syndrome with webbed neck, short stature and hypertelorism. Mentally she was normal although no formal testing was performed. There were multiple cranial nerve palsies involving left 3rd, 4th 5th, 6th, 7th, 8th, 9th and 12th nerve. Nasal endoscopy showed a mass in the nasopharynx and punch biopsy report was a papillary adenocarcinoma (figure 1). CT scans

showed a huge mass in the left post nasal space obliterating the left fossa of Rosenmuller. Mass extends into left parapharyngeal space eroding the left pterygoid plate. It also extends superiorly into the middle cranial fossa destructing the base of skull. (Figure 2)

Other investigations were negative including thyroid scan, CT scan of the chest abdomen and pelvis which showed absent uterus and ovaries in keeping with Turner's syndrome.

She had radical radiotherapy receiving a total of 60 Gray in 30 fractions over 6 weeks. Her neck had 50 Gray in 25 fractions over 5 weeks.

A repeat CT scan done 8 months (Figure 3) later showed very slight if any shrinkage of tumour. Her last follow up was 15 months after the radical therapy and she still demonstrates the signs at presentation without deterioration and she remained well.

Discussion

Papillary adenocarcinoma is uncommon in the nasopharynx and is thought to arise from the minor salivary glands. There are no known aetiological factors and sexual ratio appears equal although a ratio of 3 female to 1 male had been quoted². By and large the most common nasopharyngeal neoplasm are the WHO type I-III carcinomas while lymphomas, sarcoma,

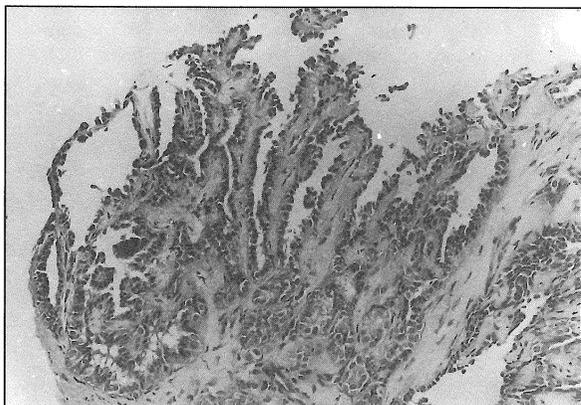


Fig. 1: Adenocarcinoma, composed of many papillary structures. (Hematoxylin & Eosin x 80)

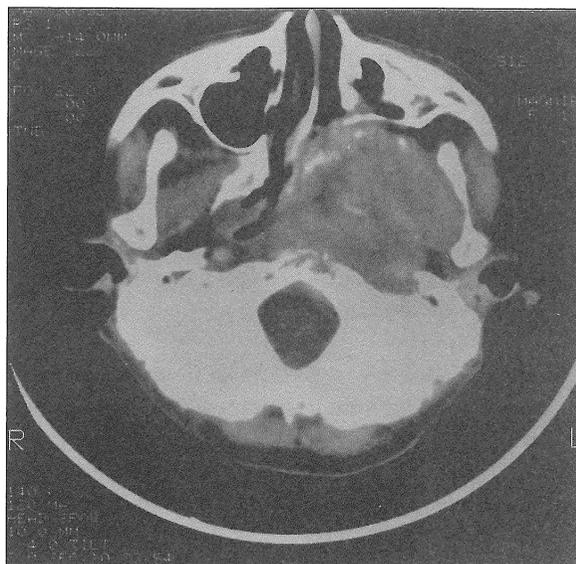


Fig. 2: Mass in the left post nasal space before radiotherapy

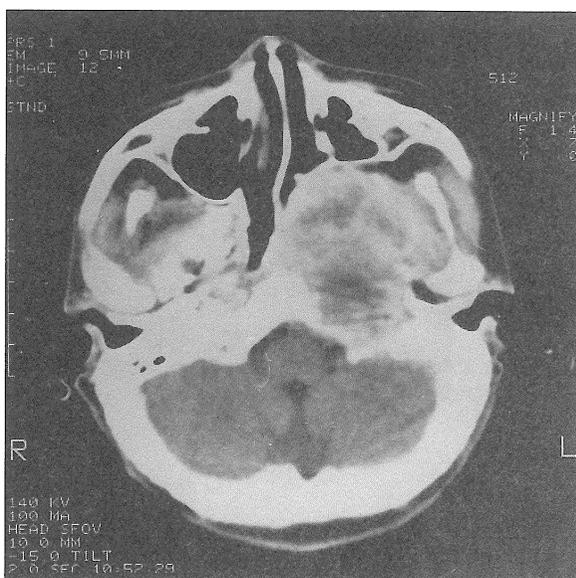


Fig. 3: Mass in the left post nasal space 8 months after radiotherapy

angiofibroma and minor salivary gland tumours make up the rest.

Weing *et al*³ reported a retrospective study on 9 patients who were diagnosed to have low grade

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nasopharyngeal adenocarcinoma. They all underwent transpalatal resection, two of them followed by postoperative radiotherapy and one had primary radiotherapy but because of persistence of tumour he also subsequently had resection. Their local disease appeared controlled with follow up duration ranging from 1-14 years. This suggest that their prognosis is good provided that they are treated aggressively. Since then there had been case reports^{1,2} that supports the strategy of surgery with equally good outcome.

The role of primary radiotherapy in nasopharyngeal adenocarcinoma or minor salivary gland tumours has not been fully evaluated. In Wernig *et al's*³ paper one patient had primary radiotherapy but was deemed a failure because of persistence rather than progression of disease. In the absence of any data it seems logical to adhere to surgery first followed by radiotherapy only if indicated.

This patient had primary radical radiotherapy as tumour had invaded the brain stem with multiple

cranial nerves palsy that surgery was deemed inappropriate. She had been followed up for 15 months and tumour was demonstrated to persist though symptomatically she had not progressed supporting the good prognosis as demonstrated by Wernig³. It would be useful to compare radiotherapy against surgery but in reality this would be extremely difficult owing to the rarity of this tumour. Until and if that ever happens all such tumours should be excised if possible.

The undocumented association between Turner's syndrome and nasopharyngeal papillary adenocarcinoma merit special mention. Turner's syndrome is associated with a number of malignancy mainly gynaecological in type⁴. To our knowledge papillary adenocarcinoma of the nasopharynx has not been reported in patients with Turner's syndrome and this would be the first such case. Perusal of the literature through Cancerlit and Medline did not yield any report of such association.

References

1. Van Hasselt CA, Ng HK. Papillary Adenocarcinoma of the Nasopharynx. *The Journal of Laryngology and Otology* 1991;105 : 853-4.
2. Wernig BM, Harpaz N, Delbridge C. Polymorphous Low-Grade Adenocarcinoma of Seromucous Glands of the Nasopharynx. *Am J Clin Pathol* 1989;92 : 104-9.
3. Wernig BM, Hyams VJ, Heffner DK. Nasopharyngeal Papillary Adenocarcinoma: A Clinicopathologic Study of a Low-Grade Carcinoma. *Am J Surg Pathol* 1988;12(12) : 946-53.
4. Ono T, Hayashi N, Saito M, Kawagoe S, Hiroi M. 45 XO/46 X dic (Yq) mosaicism in Turner's phenotype with endodermal sinus tumour of the ovary. *Gynaecol Obstret. Invest* 1989;27 : 45-7.