Plexiform Neurofibroma of the Submandibular Salivary Gland: A Rare Tumour

Shekar Y. Tati • Gautam N. Gole • Shailaja Prabhala •
Sheetal G. Gole

Received: 20 January 2010 / Accepted: 28 October 2010 / Published online: 18 November 2010
© Association of Surgeons of India 2010

Abstract A 15 year old boy presented with swelling in the submandibular region. X ray of the part showed faint radio opaque shadow. A provisional diagnosis of sialadenitis with sialolithiasis was made. Excised mass was reported histopathologically as plexiform neurofibroma of submandibular salivary gland. Plexiform neurofibroma of the salivary gland is a rare benign tumour, often present in the parotid gland. It is very rare in submandibular salivary gland. It is a slow growing, locally infiltrating tumour.

Keywords Submandibular salivary gland • Plexiform neurofibroma • Sialadenitis • Sialolithiasis

Electronic supplementary material The online version of this article (doi:10.1007/s12262-010-0174-5) contains supplementary material, which is available to authorized users.

S. Y. Tati • G. N. Gole
Department of General Surgery,
Kamineni Institute of Medical Sciences,
Sreepuram, Narketpally,
Dist - Nalgonda, Andhra Pradesh, India

S. Prabhala • S. G. Gole
Department of Pathology, Kamineni Institute of Medical Sciences,
Sreepuram, Narketpally,
Dist - Nalgonda, Andhra Pradesh, India

G. N. Gole (✉)
D/111/10, Family Quarters,
Kamineni Institute Of Medical Sciences,
Sreepuram, A/P – Narketpally,
Dist – Nalgonda, Andhra Pradesh PIN - 508254, India
e-mail: drgautamgole@rediffmail.com

Introduction

Neurofibromas constitute only 0.4 % of all salivary neoplasms. Plexiform neurofibromas of the salivary glands are rare, often presenting in the parotid gland. They are very rare in submandibular salivary gland [1, 2]. They are slow growing, locally infiltrating tumours. Only 5 cases are reported so far in the literature [3]. Probably this is 6th one. We are presenting this case because of its rarity of presentation.

Case Report

A 15 year old boy was admitted with a swelling in the submandibular region, which rapidly progressed in size. There was no history of pain or increase in size of swelling while taking meal. On examination, a firm swelling in the submandibular region of size 8×5 cm, with ovoid shape and well defined borders was present. It was bidigitally palpable. No café-au-lait spots were present over the body. A faint radio opaque shadow, suggestive of calculus was noticed in the region of the gland. Provisional diagnosis was chronic sialadenitis with sialolithiasis. FNAC was reported as chronic sialadenitis.

Intraoperatively mass was arising from submandibular salivary gland. A tubular growth was extending from the mass into the adjacent tissue.

On gross examination specimen was gelatinous and vaguely nodular with greyish white to greyish yellow areas. No calculus was found in the specimen. Microscopic examination revealed a cellular tumour tissue arranged in fascicles. Tumour tissue was composed of wavy, spindly

Fig. 1 H & E section (10×) shows salivary gland tissue with interspersed, thickened nerve bundles

Schwann cells having elongated nuclei with eosinophilic cytoplasmic extensions. There was abundant fibroblastic proliferation. Centrally situated nerve bundles were surrounded by neurofibroma tissue associated with faintly, loose mucoid matrix containing abundant wavy collagen fibres. No atypia was noted. Adjacent salivary gland also showed thickened nodular nerve bundles between salivary lobules. Histological features were suggestive of plexiform neurofibroma of the submandibular salivary gland (Figs. 1 & 2).

Discussion

Neurofibromas are benign nerve sheath tumours, which present in three forms; local discrete, generalized neurofibromatosis and plexiform neurofibromas. Common sites of occurrence of plexiform neurofibroma are fifth cranial nerve and extremities. Neurofibromas constitute only 0.4 % of all

Fig. 2 H & E section (10×) shows markedly enlarged nerve segment and thickened nerve bundles within the salivary gland

Table 1 Comparative data of 5 reported cases of plexiform neurofibroma

SR. NO	AUTHOR	YEAR	REFERENCE NO.	AGE OF PATIENT (YEARS)	SEX	PRESENTING SYMPTOM	SURGERY	PATHOLOGICAL TYPE
1	Bourgeois J. M. et al	2001	1	3	Male	Submandibular mass with multiple neurofibromatosis	Excision of submandibular mass	Plexiform neurofibroma
2	Weitzner et al	1980	2	3	Female	Submandibular mass with multiple neurofibromatosis	Excision of submandibular mass	Plexiform neurofibroma
3	Tsutsumi et al	1996	5	29	Female	Submandibular mass	Excision of submandibular mass	Plexiform neurofibroma
4	Derekoy S, Sefali M	2000	9	21	Male	Submandibular mass with multiple neurofibromatosis	Excision of submandibular mass	Plexiform neurofibroma
5	M. Aribandi, W.E. Wood, D.M. Elston & D.L Weiss	2006	3	6	Female	Submandibular mass with multiple neurofibromatosis	Excision of submandibular mass	Plexiform neurofibroma

salivary neoplasms. Plexiform neurofibromas of the salivary glands are rare, usually presenting in the parotid gland. They are very rare in submandibular salivary gland.

Plexiform neurofibromas are diffuse enlargements of multiple fascicles of the nerves and its branches, leading to thickening of nerves. They grow along nerves extending into the surrounding tissue. They are slow growing and locally infiltrating benign tumours. When located deeply they may have greater chance of malignancy. Large tumours may be associated with pressure symptoms on trachea, pharynx. Sometimes they may extend into spinal canal and compress the spinal cord. They grow rapidly in children, during puberty and in pregnancy.

Plexiform neurofibromas are frequently seen in patients with type I neurofibromatosis and undergo malignant changes in 2 % of the cases. Patient with plexiform neurofibroma without family history or without features of type I neurofibromatosis require genetic work up. In such cases tumour may present as a result of local somatic mutation and patient may not transmit the disease to their offspring—[4].

Neurofibromas have been reported in parotid glands—Seifert et al [5, 6]. Out of 300 neurofibromas in the minor salivary gland tumours, two were found to be plexiform neurofibromas [7]. Plexiform neurofibromas are exceedingly rare in submandibular and sublingual salivary glands [1, 2, 8, 9].

A search in the literature revealed only 5 cases of plexiform neurofibromas of submandibular salivary gland {please see Table 1}.

Out of 5 cases 3 were females. 4 out of above 5 cases presented with type I neurofibromatosis. However in our case there was no type I stigma present.

If we consider FNAC to diagnose plexiform neurofibroma, its diagnostic yield appears to be low. A plexiform neurofibroma may be suspected in the appropriate setting, with the help of cross sectional imaging. CT imaging may show conglomerate multilobulated masses that may appear as a ‘bag of worms’. There is hypoattenuation with heterogenous contrast enhancement. A characteristic “branching” hypoattenuated mass on CT, with branching tubular masses extending into adjacent tissue could virtually be diagnostic [10]. In our case CT was not done.

Histopathologically there is diffuse cylindrical enlargement of multiple fascicles of the nerve and its branches. There is a myxoid matrix containing Schwann cells, nerve fibers, mast cells and perineurial endoneurial fibroblasts.

Surgical excision is the treatment of choice. Recurrence may occur. It is reported in 20 % of the patients with plexiform neurofibroma after complete resection and increases to 44 % with incomplete resection (Where there is involvement of vital structures) [3].

Acknowledgement To our Principal (KIMS), Director (KES) and management.

Presentation in conference Not presented

Financial support Nil

Conflict of interest Nil.

References

1. Bourgeois JM, Radhi J, denEi L et al (2001) Plexiform neurofibromas of the submandibular gland in a child. *Can J Gastroenterol* 15:835–837
2. Weitzner S (1980) Plexiform neurofibromas of the submandibular gland in children. *Oral Surg Oral Med Oral Pathol* 50:53–57
3. Aribandi M, Wood WE, Elston DM, Weiss DL (2006) CT features of plexiform neurofibroma of the submandibular gland. *Am J Neuroradiol* 27:126–128
4. Scheithaur BW, Woodruff JM, Erlandson RA (1999) Tumours of the peripheral nervous system. Armed Force Institute of Pathology, Washington
5. Komat TT, Suzaki A (1996) Solitary plexiform neurofibromas of the submandibular gland. *J Laryngol Otol* 110:1173–75
6. Seifert G, Meihike A, Haubrich J et al (1986) Pathology, diagnosis, treatment, facial nerve surgery, Stuttgart Georg Thieme Verlag 171–301
7. Huvos CEB, AG SEW et al (1972) Tumours of the major salivary glands in children. *Cancer* 29:312–317
8. Kahwaji G, Hamdan AL, Mufarij A et al (2000) Plexiform neurofibromas of the submandibular gland. *Otolaryngol Head Neck Surg* 122:1927
9. Derekoy S, Sefali M (2000) Plexiform neurofibromas of the submandibular gland. *J Laryngol Otol* 114:643–645
10. Lin J, Martel W (2001) Cross sectional image of peripheral nerve sheath tumours: characteristic sign on CT, MR imaging and sonography. *AJR Am J Roentgenol* 176:75–82