Neurofibroma of External Ear: The Updates

Dear Editor,

I would like to resubmit this article for the kind consideration for publication in your esteemed journal. I am happy to inform you that I have thoroughly gone through the literature to address the issues raised by the reviewers with evidences. I give a point wise reply to the reviewers' comments.

Reviewers' Comments

1. Which histopathologic variant is common among external ear neurofibroma (NF)?

   Of three histopathological types, that is, localized, plexiform, and diffuse;[1] diffuse variant involves skin and subcutaneous tissues of head neck region.[2-6] Among head neck region, periauricular area is the most common site.[1] So diffuse variant is common among external ear NF.
2. How long the patients need follow-up?

   Follow-up is mainly to identify an early recurrence in operated cases. If we are planning observation only, wait and watch at regular intervals with radiological evaluation and radical excision if there is an evidence of tumor growth.[7]

   As we know that diffuse NFs recur frequently.[8] Follow-up should depend on the age of the patient, extent and location of tumor, and partial or complete excision status.

   We can expect a less recurrence and hence increase the follow-up duration when there is a gross total resection and low residual tumor volume.[9] Proper counseling regarding early follow-up is needed, if there is rapidly enlarging mass, neurogenic pain/motor weakness, or disfiguring.[10]

   Literature does not mention the exact duration for follow-up. van Zuuren and Posma[2] advised yearly follow-up in their article.
3. Is magnetic resonance imaging (MRI) necessary in all these patients?

   Imaging is done to know the characteristic, extent, and deeper extension of a soft tissue lesion. Although NFs are superficial lesions, which can be evaluated clinically, but it is always better to carry out radiology before histopathological confirmation. Ultrasonography (USG) and computed tomography (CT) can assess the nature but cannot demarcate between the vascularity of the lesion. USG has the advantages of no radiation exposure, cost-effectiveness, and early reports; it also carries limitations of not assessing the extent and depth of larger lesions. CT can help to know the extent when bony external auditory canal starts getting involved. MRI is the investigation of choice as it demarcates the tumor with the surrounding structures; it also differentiates between all three varieties, that is, local, plexiform, and diffuse. USG and CT scans are less reliable as diffuse NF resembles lipoma or hemangioma.[11]

   If MRI suggests a highly vascular lesion, consider a preoperative angiogram, and if necessary, a preoperative intra-arterial embolization, if hemorrhage is anticipated.[12]

   Therefore, in resourceful settings, we should consider MRI as a necessary investigation in these patients.
4. What are the long-term complications of surgery in these patients?

   The long-term complications of surgery are recurrence, hypertrophic scarring,[13] permanent neurological deficit, functional impairment, wound healing abnormalities,[14,15] and rarely malignant transformation.[16,17]

   Age <10 years at surgery, head-neck-face-trunk lesion, and incomplete resection take shorter relapse time.[9,18]

   Several authors advised for wide meatoplasty to prevent reobstruction.[19] We have a case that reports wide external auditory canal after 5.5-year follow-up.[20]

   Therefore, the major concerns to be kept in mind during surgery are the extent of resection in balance to the likelihood of recurrence and loss of function.

Acknowledgement

I thank the reviewers for the helpful suggestions and for the prudent handling of my article.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

References

1. Megehed M. Histopathological variations of neurofibroma. A study of 114 lesions. Am J Dermatopathol 1994;16:486-95.
2. van Zuuren EJ, Posma AN. Diffuse neurofibroma on the lower back. J Am Acad Dermatol 2003;48:938-40.
3. Kapadia SB, Janecka IP, Curtin HD, Johnson BL. Diffuse neurofibroma of the orbit associated with temporal meningocele and neurofibromatosis-1. Otolaryngol Head Neck Surg 1998;119:652-5.

4. de Varebeke SJ, De Schepper A, Hauben E, Declau F, Van Marck E, Van de Heyning PH. Subcutaneous diffuse neurofibroma of the neck: a case report. J Laryngol Otol 1996;110:182-4.
5. Enzinger M, Weiss SW, Goldblum JR. Benign tumors of peripheral nerves. In: Enzinger and Weiss's soft tissue tumors. 4th ed. St. Louis, MO: Mosby; 2001. pp. 1132-40.
6. Kransdorf MJ, Murphey MD. Neurogenic tumors. In: Imaging of soft tissue tumors. 2nd ed. Philadelphia, PA: Lippincott Williams & Wilkins; 2006. pp. 334-8.
7. Coakley D, Atlas MD. Diffuse neurofibroma obstructing the external auditory meatus. J Laryngol Otol 1997;111:145-7.
8. Trevisani TP, Pohl AL, Matloub HS. Neurofibroma of the ear: function and aesthetics. Plast Reconstr Surg 1982;70:217-9.
9. Needle MN, Cnaan A, Dattilo J, Chatten J, Phillips PC, Shochat S, *et al*. Prognostic signs in the surgical management of plexiform neurofibroma: the Children's Hospital of Philadelphia experience, 1974–1994. J Pediatr 1997;131:678-82.
10. Wise JB, Cryer JE, Belasco JB, Jacobs I, Elden L. Management of head and neck plexiform neurofibromas in pediatric patients with neurofibromatosis type 1. Arch Otolaryngol Head Neck Surg 2005;131:712-8.
11. Hassell DS, Bancroft LW, Kransdorf MJ, Peterson JJ, Berquist TH, Murphey MD, *et al*. Imaging appearance of diffuse neurofibroma. AJR Am J Roentgenol 2008;190:582-8.
12. Ergu¨n SS, Emel E, Karabekir S, Bu¨yu¨ kbabani N. Extracranial diffuse neurofibroma with intracranial extension. Plast Reconstr Surg 2000;105:801-3.
13. Ferner RE, Huson SM, Thomas N, Moss C, Willshaw H, Evans DG, *et al*. Guidelines for the diagnosis and management of individuals with neurofibromatosis 1. J Med Genet 2007;44:81-8.
14. Prada CE, Rangwala FA, Martin LJ, Lovell AM, Saal HM, Schorry EK, *et al*. Pediatric plexiform neurofibromas: impact on morbidity and mortality in neurofibromatosis type 1. J Pediatr 2012;160:461-7.
15. Canavese F, Krajbich JI. Resection of plexiform neurofibromas in children with neurofibromatosis type 1. J Pediatr Orthop 2011;31:303-11.
16. Crowe FW, Schull WJ, Neel JV. Multiple neurofibromatosis. Springfield, Illinois: Charles C Thomas; 1956.
17. Feinman NL, Yakomac BA. Neurofibromatosis in childhood. J Pediatr 1970;76:339.
18. Donner TR, Voorhies RM, Kline DG. Neural sheath tumors of major nerves. J Neurosurg 1994;81:362-73.
19. Rombout J, van Rijn PM. M-meatoplasty: results and patient satisfaction in 125 patients (199 ears). Otol Neurotol 2001;22:457-60.
20. Minoda R, Ise M, Murakami D, Kumai Y, Yumoto E. Surgical removal of diffuse-type neurofibroma involving the auditory external canal in a patient with neurofibromatosis type 1. Int Adv Otol 2012;8:497-502.

Neha Shakrawal

Department of Otorhinolaryngology, All India Institute of Medical Sciences, Jodhpur, Rajasthan, India

Address for correspondence: Dr. Neha Shakrawal, Department of Otorhinolaryngology, All India Institute of Medical Sciences, Jodhpur 302005, Rajasthan, India.
E-mail: drnehasnmc@gmail.com

Access this article online

Quick Response Code:

Website: www.jcasonline.com

DOI: 10.4103/JCAS.JCAS_74_20

How to cite this article: Shakrawal N. Neurofibroma of external ear: The updates. J Cutan Aesthet Surg 2020;13:372-3.