Review Article

International Journal of Neurology and Neurosurgery
Volume 15 Number 2, April – June 2023
DOI: http://dx.doi.org/10.21088/ijnns.0975.0223.15223.3

Scalp Tumors: An Overview of 2 Case Reports in Our Tertiary Care Government Hospital

Agraj Mishra

How to cite this article:
Agraj Mishra/Scalp Tumors: An Overview of 2 Case Reports in Our Tertiary Care Government Hospital/Int J Neurol Neurosurg.2023;15(2): 57-61.

Abstract

Scalp tumors are among the most commonly encountered tumors. Behaviour and management of scalp tumors depends heavily on cell lineage. Appropriate identification of these tumors is paramount in mitigating disease burden. Most tumors require surgical resection at a minimum; others will require adjuvant therapies.

Keywords: Scalp tumors; Plexiform Neurofibroma; Scalp Metastasis; Adenocarcinoma; Lung Carcinoma.

INTRODUCTION

Scalp tumors form a diverse grouping of maladies, each with varying gradations of seriousness and the need for more aggressive intervention. Anatomically, the scalp is defined by the supraorbital ridges anteriorly, the superior nuchal line posteriorly, and the zygoma and mastoid inferiorly. The lymph drains via the parotid gland into the jugular and digastric nodes for the anterior scalp and via the posterior triangle of the neck for the posterior scalp. The scalp itself consists of five distinct layers: skin, connective tissue, aponeurosis (galea or epicranium), loose connective tissue, and periosteum. Within the skin layer are adnexal structures, including eccrine, apocrine, and sebaceous glands as well as hair follicles. Any of these structures may become the site of tumors, either benign or malignant. Fortunately, the majority of these tumors are benign in nature. Risk factors for scalp tumors are multifactorial and include both genetic and environmental components, as well as gene environment interactions. Environmental variables associated with increased risk include cumulative exposure to sun, radiation, or toxins. While only approximately 1–2% of all scalp tumors are malignant, they comprise up to 13% of all malignant cutaneous neoplasms.[1]

Cutaneous metastasis from a primary visceral malignancy is an uncommon entity, with an incidence ranging from 0.2%–10%.[2,3] It may occur due to direct extension of the tumor as a local metastasis or as a distant metastasis.[4] Generally, cutaneous metastasis develops after initial diagnosis of the primary internal malignancy and later in the course of the disease. In very rare cases, it may occur at the same time or before the primary cancer has been detected.[5] Skin metastasis from lung cancer is a rare clinical entity that has been reported to occur in 0.22%–12% of patients with lung cancer.[2,3,6] Skin metastasis as the initial and sole manifestation of an

Author Affiliation: Senior Resident, Department Neuro Surgery, B.J. Medical College and Sassoon Hospital, Pune 411001, Maharashtra, India.

Corresponding Author: Agraj Mishra, Senior Resident, Depatment Neuro Surgery, B.J. Medical College and Sassoon Hospital, Pune 411001, Maharashtra, India.

E-mail: mishraagraj37@gmail.com

Received on: 06.04.2023

Accepted on: 01.05.2023

© Red Flower Publication Pvt. Ltd.

underlying lung cancer is a very rare occurrence.[7] Most common sites of skin metastases from lung cancer are the chest, abdomen, head and neck. They may rarely appear in the form of solitary lesion on the scalp.[8] Compared to other malignancies, lung cancer is the fastest in developing skin metastases after initial diagnosis.[9] Cutaneous metastasis of lung cancer is physically indistinguishable from those due to carcinoma originating elsewhere in the body. Lung cancer should always be considered in the differential diagnosis of patients with nodular skin lesions. With a very poor prognosis, the median survival time after the diagnosis of the cutaneous metastasis is between 2.9 – 4.9 months.[10]

Neurofibroma is a benign tumor of the peripheral nerves heath. It arises due to the abnormal proliferation of Schwann cells, perineural cells, and endoneural fibroblasts. Several types of pathology are identified, including localized, plexiform, and diffuse types.[11,12] Neurofibromatosis type 1 (NF1) is a neurocutaneous, autosomal dominant disorder.[13] The disease manifestations are café-au-lait macules (CALMs), neurofibromas, skin folding freckling, iris hamartomas (Lisch's nodules), optic pathway gliomas, and skeletal deformities.[14,15] Plexiform neurofibroma is a benign tumor of the peripheral nerves (World Health Organization grade I) and an unusual variant of neurofibroma that arises from a proliferation of all parts of the nerve. Plexiform neurofibroma is essentially pathognomonic of NF1.[16-18] This tumor has a significant risk of eventual malignant transformation.[17] The possibilities of malignancy and recurrence are the main reasons for long term, close follow-up.[16-23]

Case Report 1: A 70 years old female presented with scalp swelling on left frontal region above hair line present since last 8 month and increases rapidly and associated with no pain. She had no history of lung disease. She denied any respiratory symptoms, fever, or weight loss, and his general condition was good. Her blood results were within normal limits.



Fig. 1 & 2: Picture showing swelling in right frontal region and post-operative picture showing scalp reconstruction.

Clinical examination revealed a painless, movable, nonulcerated nodule in scalp measuring approximately 5.0 × 4.0 cm. The overlying skin was red due to enlarged mass. On radiological investigation, the swelling was confined to scalp and not penetrated the vault or dura. Further investigations with chest X-ray show mass in lower lobe of right lung. Subsequent computed tomography (CT) showed a large mass involving the right lower lobe.

After careful planning and evaluation, excision of mass along with overlying skin reconstruction with rotational flap along split thickness skin graft performed.

On histopathology examination, confirmed the presence of adenocarcinoma lung and positive for periodic acid Schiff (PAS). She was advised to start chemotherapy and radiotherapy. Unfortunately, there were no improvements on either the cutaneous metastatic mass or her general condition.

Case report 2: A 17 years old boy presented with scalp swelling for past 6 years located in right occipital region and was painless. He had many cutaneous neurofibroma on chest, forearm and legs. Clinical examination revealed a soft boggy mass in occipital region which of size 6*4 CMS and not fixed to underlying structure. On radiological evaluation, demonstrated a mass in the posterior scalp, right parietooccipital region and the underlying bone shows hyperostosis. Patient presented with CALMs in the back region with a variation of diameters.



Fig. 3 & 4: A 17 years old boy showing swelling at occipital region and picture at the time of resection in operation theatre.

Resection of the mass was performed along with skin reconstruction. The anatomical pathology result was plexiform neurofibroma. And patient was discharged with advice to follow up.

DISCUSSIONS

Dissemination of visceral malignancies to the skin is rather rare and usually occurs in a later stage of the disease. However, cutaneous metastases may be the first indication of the clinically silent visceral malignancies. In case of lung cancer, metastasis to the skin is much less frequent than that to other organs (brain, bone, liver and adrenal glands). In a recent meta-analysis of six studies containing over 20 000 patients with cutaneous metastasis, the overall incidence from all visceral malignancies was estimated to be 5.3%.[24] A retrospective study in 2012 indicated that 2.8% of 2130 patients with advanced non-small cell lung cancer (NSCLC) showed cutaneous metastases as an initial presentation.[25] In a retrospective study by Lookingbill *et al*. including 7316 cancer patients, skin involvement as a presenting sign was seen in only 0.8%. In literature, various data are reported related to the frequency of skin metastases of different histological types of lung cancer. Dreizen *et al*.[26] reported that adenocarcinoma has the highest tendency to metastasize to skin. Brownstein and Helwing[27] reported that adenocarcinoma and squamous cell carcinoma show the equal tendency to involve the skin; while Terashima and Kanazawa and Hidaka *et al*.[26] noted that the cutaneous metastasis rate was high for large cell carcinomas and low for squamous and small cell variants. Therefore, the histological type of lung cancer with the highest incidence of cutaneous metastases seems to be debated yet. Physically, cutaneous metastatic lesions due to lung cancer are indistinguishable from those due to carcinoma originating else where in the body. They may arise from the pilosebaceous unit, from the interfollicular epidermis, or dermis. Cutaneous metastasis can manifest as a nodule, ulceration, cellulitis like lesion, bullae or fibrotic process. Generally, the nodular type is the result of hematogenous metastasis, and is likely to be the most common. Nodules are painless, mobile or fixed, firm or rubbery, discrete or multiple. They vary in colour from flesh tones to red-purple, or blue-black and vary in diameter from 5 mm to 6 cm. Multiple lesions are usually grouped. They initially grow rapidly, and then more slowly, and may necrotize or ulcerate.[26] Chiu *et al*. reviewed the data of 398 patients with malignant scalp tumors and found that the basal and squamous cell carcinomas were the most common histologic subtypes.[28] Scalp metastasis as the initial manifestation of an underlying lung cancer is an exceedingly rare clinical entity. In most cases the lesions are multiple.[26] In our patient, clinical presentation was very uncommon, since the solitary nodule was movable and the overlying skin was intact. Increased awareness is needed, as a similar clinical presentation can be attributed to an epidermoid or a trichilemmal cyst, especially when the patient is completely asymptomatic. Epidermoid and trichilemmal cysts are the most common causes of solitary scalp nodules in adults.[29] These patients have an extremely poor prognosis with an average survival ranging between three and five months in the majority of studies.[29] Patients that present with skin metastases earlier during the disease course, have poorer prognosis compared to those with later developed metastases. Miyazaki *et al.* reported a case of spontaneous regression of scalp metastases from lung cancer.[30]

NF1 is a condition of rare autosomal dominant genetic mutations of the NF1 gene at chromosome 17q11.2.[31] Clinical manifestations of this abnormality consist of multiple skin alterations such as CALMs and axillary freckling, and by tumoral growth along nerves, called "neurofibromas."[31,35,45] Plexiform neurofibroma is an unusual type of neurofibroma and a benign tumor of the peripheral nerve that possesses a high risk of malignant transformation.[35,43,46,48] Plexiform neurofibromas are usually diagnosed in early childhood and found in approximately 30% of NF1 cases, most frequently in the craniomaxillofacial region.[42,44] Malignant transformation occurs in 2% to 16% of cases and is considered the leading cause of mortality.[47] NF1 is diagnosed using 2 or more criteria from the National Institutes of Health. Plexiform neurofibromas are diagnosed clinically by the typical manifestations of the disease. Histopathological studies are performed to differentiate malignant change.[43-48] Our patient had the manifestations of numerous CALMs, pseudoarthrosis, cutaneous neurofibroma, and one plexiform neurofibroma. Surgery remains the treatment of choice in plexiform neurofibromas. Total resection of the tumor mass is performed when malignant transformation occurs, especially in recurrent cases. The incidence of recurrence is 20% of cases, even when the approach was done appropriately.[41,42] Given this concern and its nature, some authors[43,44] Suggest that neurofibroma should receive a more radical tumor mass resection. Even this tumor recurrence may occur even if completely resected. The tumor resection aims to relieve the symptoms.[44–46] This patient had undergone total resection of the tumor. Four years later, the patient presented with a recurrent tumor in the same location as previously and a pain that did not

subside with nonopioid medication.

CONCLUSION

Solitary scalp metastasis as the first sign of an occult non-small cell lung cancer is an extremely rare occurrence. Despite its rarity, metastatic skin disease should always be considered in the differential diagnosis in patients with a history of smoking or lung cancer presenting with cutaneous nodules. Increased awareness of this rare entity is needed for early recognition and initiation of the appropriate treatment. More over, it helps in appropriate staging, altering therapy and a better estimation of prognosis. We also conclude that a cutaneous metastasis is the most common dermatological manifestation of lung cancer.

Plexiform neurofibromas exhibit an unusual type of NF1 with a higher risk of recurrence after surgical procedure. Recurring tumor harbours a high risk for malignant transformation. Accomplishing a good outcome can be related to good perioperative planning and precise operative procedure, without local recurrence complications. Periodic clinical and radiology imaging examinations are essential in the evaluation of the recurrence.

Disclosures

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

REFERENCES

1. Scalp Tumors J. Scott Persing, Sarah M. Persing, James E. Clune, and John A. Persing. *Youman's and winn 7 edition*.
2. Lee J.H., Ahn S.J., Kim H.J., Jang S.E., Noh G.Y., Kim H.R. Cutaneous metastasis from lung cancer: a single-institution retrospective analysis. Tuberc. *Respir. Dis*. 2011;70(2):139–142.
3. Song Z., Lin B., Shao L., Zhang Y. Cutaneous metastasis as an initial presentation in advanced non-small cell lung cancer and its poor survival prognosis. J. Cancer Res. Clin. *Oncol*. 2012;138(10):1613–1617.
4. Looking bill D.P., Spangler N., Sexton F.M. Skin involvement as the presenting sign of internal carcinoma. A retrospective study of 7316 cancer patients. J. Am. Acad. *Dermatol*. 1990;22(1):19–26.
5. Mollet T.W., Garcia C.A., Koester G. Skin metastases from lung cancer. Dermatol. *Online J*. 2009;15:1. (PubMed).
6. Molina Garrido M.J., Guilĺen Ponce C., Soto Marınez J.L., Marınez Y., Sevila C., Carrato Mena A. Cutaneous metastases of lung cancer. *Clin. Transl. Oncol*. 2006;8(5):330–333.
7. Hidaka T., Ishii Y., Kitamura S. Clinical features of skin metastasis from lung cancer. *Intern. Med*. 1996;35:459–462.
8. Kikuchi Y., Matsuyama A., Nomura K. Zosteriform metastatic skin cancer: report of three cases and review of the literature. *Dermatology*. 2001;202:336–338. (PubMed).
9. Marcoval J., Moreno A., Peyrí J. Cutaneous infiltration by cancer. *J. Am. Acad. Dermatol*. 2007;57:577–580. (PubMed).
10. Perng D.W., Chen C.H., Lee Y.C., Perng R.P. Cutaneous metastasis of lung cancer: an ominous prognostic sign. *Zhonghua Yi Xue Za Zhi (Taipei)* 1996;57:343–347.
11. Albright AL, Pollack IF, Adelson PD. Principles and Practice of Pediatric Neurosurgery. 3rd ed. New York, NY: *Thieme publishers*; 2015.
12. Tonsgard JH. Clinical manifestations and management of neurofibromatosis type 1. *Semin Pediatr Neurol*. 2006;13(1):2–7.
13. Jallo GI, Kothbauer KF, Recinos VMR. Handbook of Pediatric Neurosurgery. New York, NY: *Thieme publishers*; 2018.
14. Krohel GB, Rosenberg PN, Wright JE, *et al.* Localized orbital neurofibromas. *Am J Ophthalmol*. 1985;100(3):458–464.
15. Ardashev VN, Seriakov AP, Nikolaeva SN, *et al.* Diagnostics of neurofibromatosis (Recklinghausen disease). Article in Russian. *Voen Med Zh*. 2004;325(6):41–44.
16. Ferner RE, Huson SM, Thomas N, *et al.* Guidelines for the diagnosis and management of individuals with neurofibromatosis 1. *J Med Genet*. 2007;44(2):81–88.
17. Prada CE, Rangwala FA, Martin LJ, et al.. Pediatric plexiform neurofibromas: impact on morbidity and mortality in neurofibromatosis type 1. *J Pediatr*. 2012;160(3):461–467.
18. Raja AI, Yeaney GA, Jakacki RI, *et al.* Extraventricular neurocytoma in neurofibromatosis type 1: case report. *J Neurosurg Pediatr*. 2008;2(1):63–67.
19. Yohay K. Neurofibromatosis type 1 and associated malignancies. *Curr Neurol Neurosci Rep*. 2009;9(3):247–253.
20. Matrix H, Origin U, Laffan EE, *et al.* Pediatric soft-tissue tumors and pseudotumors : MR imaging features with pathologic correlation. *Radiographics*. 2009;36:1–35.

21. Weiss LM. Soft Tissues. 2nd ed. *Elsevier Inc.*; 2009.
22. Jett K, Friedman JM. Clinical and genetic aspects of neurofibromatosis 1. *Genet Med.* 2010;12(1):1–11.
23. Abbas O, Bhawan J. Cutaneous plexiform lesions. *J Cutan Pathol.* 2010;37(6):613–623.
24. Krathen R.A., Orengo I.F., Rosen T. Cutaneous metastasis: a meta-analysis of data. *South. Med. J.* 2003;96:164–167.
25. Fay T., Henry G.C. Correlation of body segmental temperature and its relation to the location of carcinomatous metastasis: clinical observations and response to methods of refrigeration. *Surg. Gynecol. Obstet.* 1938;66:512–514.
26. 16. Dreizen S., Dhingra H.M., Chiuten D.F., Umsawasdi T., Valdivieso M. Cutaneous and subcutaneous metastases of lung cancer: clinical characteristics. *Postgrad. Med.* 1986;80:111–116.
27. 17. Browntein M.H., Helwig E.B. Metastatic tumors of the skin. *Cancer*. 1972;29:1298–1307.
28. 18. Chiu C.S., Lin C.Y., Kuo T.T., Kuan Y.Z., Chen M.J., Ho H.C. Malignant cutaneous tumors of the scalp: a study of demographic characteristics and histologic distributions of 398 Taiwanese patients. *J. Am. Acad. Dermatol*. 2007;56(3):448–452.
29. 19. Farley R., Manolidis S., Ratner D. Adenocarcinoma of the lung metastatic to the skull presenting as a scalp cyst. *J. Am. Acad. Dermatol*. 2006;54:916–917.
30. 20. Miyazaki K., Masuko H., Satoh H., Ohtsuka M. Lung cancer with spontaneous regression of scalp metastasis. *Respir. Med. Extra.* 2007;3(2):83–85.
31. Khajavi M, Khoshsirat S, Ahangarnazari L, *et al.* A brief report of plexiform neurofibroma. *Curr Probl Cancer*. 2018;42(2):256–260.
32. Tchernev G, Chokoeva AA, Patterson JW, *et al.* Plexiform neurofibroma: a case report. *Medicine (Baltimore)*. 2016;95(6):e2663.
33. Williams VC, Lucas J, Babcock MA, *et al.* Neurofibromatosis type 1 revisited. *Pediatrics.* 2009;123(1):124–133.
34. Gross AM, Singh G, Akshintala S, *et al.* Association of plexiform neurofibroma volume changes and development of clinical morbidities in neurofibromatosis 1. *Neuro Oncol.* 2018;20(12):1643–1651.
35. Bakshi SS. Plexiform neurofibroma. *Cleve Clin J Med*. 2016;83(11):792.
36. Polak M, Polak G, Brocheriou C, *et al.* Solitary neurofibroma of the mandible: case report and review of the literature. *J Oral Maxillofac Surg.* 1989;47(1):65–68.
37. Evans DG, Howard E, Giblin C, *et al.* Birth incidence and prevalence of tumor-prone syndromes: estimates from a UK family genetic register service. *Am J Med Genet A*. 2010;152A(2):327–332.
38. Liu S, Zhou X, Song A, *et al.* Giant plexiform neurofibroma of thigh in a young woman. *Postgrad Med J.* 2019;95(1126):459–460.
39. Ruggieri M, Pavone V, Polizzi A, *et al.* Unusual form of recurrent giant cell granuloma of the mandible and lower extremities in a patient with neurofibromatosis type 1. Oral Surg Oral Med *Oral Pathol Oral Radiol Endod*. 1999;87(1):67–72.
40. Sabatini C, Milani D, Menni F, *et al.* Treatment of neurofibromatosis type 1. *Curr Treat Options Neurol*. 2015;17(6):355.
41. Kumar S, Bhaskar S, Handa A, *et al.* Diffuse neurofibroma of scalp. Asian J Neurosurg. 2014;9(4):237.
42. Smith KB, Wang DL, Plotkin SR, *et al.* Appearance concerns among women with neurofibromatosis: examining sexual/bodily and social self-consciousness. *Psychooncology*. 2013;22(12):2711–2719.
43. Riccardi VM. Von Recklinghausen neurofibromatosis. *N Engl J Med*. 1981;305(27):1617–1627.
44. Fried rich RE, Schmelzle R, Hartmann M, *et al.* Resection of small plexiform neurofibromas in neurofibromatosis type 1 children. *World J SurgOncol*. 2005;3(1):6.
45. Needle MN, Cnaan A, Dattilo J, *et al.* Prognostic signs in the surgical management of plexiform neurofibroma: the Children's Hospital of Philadelphia experience, 1974-1994. *J Pediatr.* 1997;131(5):678–682.
46. Balaji SM. Surgical management of recurrent neurofibroma of infratemporal region: a case report with 20-year follow-up. *Indian J Dent Res*. 2017;28(6):695–698.
47. Roberts AHN, Crockett DJ. An operation for the treatment of cutaneous neurofibromatosis. *Br J Plast Surg*. 1985;38(2):292–293.
48. Parsons CM, Canter RJ, Khatri VP. Surgical management of neurofibromatosis. *Surg Oncol Clin N.*