

Primary Epithelioid Sarcoma of Frontotemporal Scalp: a Rare Case Report with Recent Literature Review

Adhikari Ganga D, Mandal Sandip Kumar, Gnawali Suman, Shrestha Gyan B, Sharma Nitu, *Yadav Ajay Kumar

Department of Radio-Diagnosis, Imaging and Nuclear Medicine, BP Koirala Memorial Cancer Hospital, Bharatpur, Nepal

Abstract

Epithelioid sarcoma (ES) is a very rare and aggressive mesenchymal sarcoma subtype which represents less than 1% of soft tissue sarcomas (STS). According to the origin of the site, there are two types of ES: distal-type epithelioid sarcoma and proximal-type epithelioid sarcoma. The clinical diagnosis of ES mainly is confirmed by histopathology examination followed by immunohistochemistry. Radical excisional surgery is the best treatment option for epithelioid sarcoma. According to TNM staging, the treatment option will vary from surgery to surgery accompanied with radiotherapy and chemotherapy. Here in, we presented a 36-year-old man had non-healing ulcer in left frontotemporal scalp extending to involve left orbit. Histopathological confirmed epithelioid sarcoma. Magnetic Resonance Imaging (MRI) of Head and Neck revealed that there was mass in left frontotemporal scalp measuring 6.7 X 5.7 X 2.6 cm extending to zygomatic region and left orbit D/D malignant mass/sarcoma with bilateral (B/L) sinusitis. After that patient undergone for Wide Local excision (WLE) surgery on 11th September 2022. Adjuvant Radiation Therapy (RT) 6000 cGy radiation dose in 30 fractions (#) which was 200 cGy per fraction (#) were given during 22nd September to 10th November 2022 due to local advancement of disease. Patient was asked to follow up after 6 weeks after completion of RT. After 6 weeks of surgery, the patient was undergone for MRI and report revealed normal study.

Keywords: Frontotemporal region, epithelioid sarcoma, Wide Local excision, mesenchymal sarcoma, Histopathology

Introduction

Epithelioid sarcoma (ES) is a very rare and brutal soft tissue sarcoma first portrayed by Enzinger in 1970, which may be proximal or distal types.¹ ES is a malignant mesenchymal neoplasm that exhibits epithelioid cytomorphology and a predominantly epithelial phenotype. The neoplastic cells in the proximal subtype tend to exhibit prominent malignant features including moderate nuclear atypia with prominent nucleoli and are associated

with a poorer prognosis than the distal subtype.² ES of the scalp as a primary site of origin is a rare occurrence and hence possesses a challenge in early diagnosis and timely intervention. In addition, there is not specific treatment management for ES of the scalp due to very few cases were reported and limited studies are accessible on the role of chemotherapy. Therefore, even in the current period, radical wide surgical resection (WLE) with

Correspondence: Yadav Ajay Kumar, Department of Radiodiagnosis, Imaging and Nuclear Medicine, B.P. Koirala Memorial Cancer Hospital, Bharatpur, Nepal.

adjuvant radiation therapy remains the basis of treatment.³ ES has a far above the ground predisposition for local recurrence, lymph nodes invasion, and distant metastasis, especially when it involves the lungs.⁴

Many research studies articles have reported on epithelioid sarcoma survival rates through the years. Due to rare occurrences of the disease, the numbers show a discrepancy and significantly. Five-year survival and ten-year survival rate for patients with epithelioid sarcoma are approximately 50-70% and 42-55% respectively.⁵ Our case was a survival case from ES in the scalp. There were 14 primary studies that have been reported ES in the scalp till the date. We did a retrospective analysis and reviewed all the reported studies.

Case Report

Here in, we presented a 36-year-old man had non-healing ulcer in left frontotemporal scalp extending to involve left orbit. Histopathological features and immunohistochemistry were confirmed that it was proximal type epithelioid sarcoma. Histopathology examination revealed that pleomorphic dermal infiltrate consistent within epithelioid sarcoma. Magnetic Resonance Imaging (MRI) of Head and Neck revealed that there was mass in left frontotemporal scalp measuring 6.7 X 5.7 X 2.6 cm extending to zygomatic region and left orbit D/D malignant mass/sarcoma with bilateral (B/L) sinusitis.

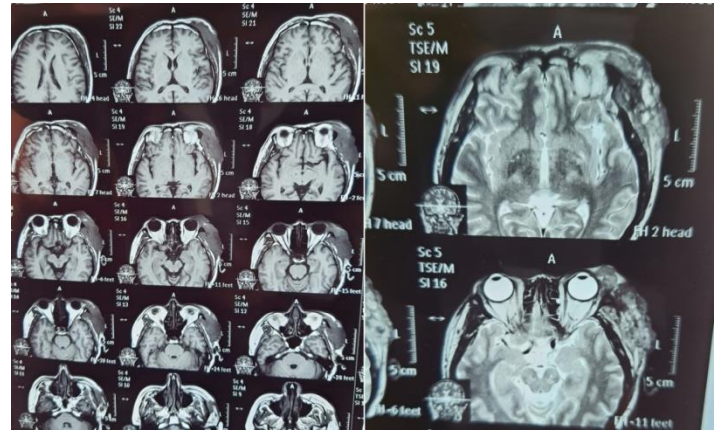


Figure 1: Pre-treated Magnetic Resonance Imaging (MRI) scan of Face and Neck revealed that there was mass in left frontotemporal scalp measuring 6.7 X 5.7 X 2.6 cm extending to zygomatic region and left orbit D/D malignant mass/sarcoma with bilateral (B/L) sinusitis.

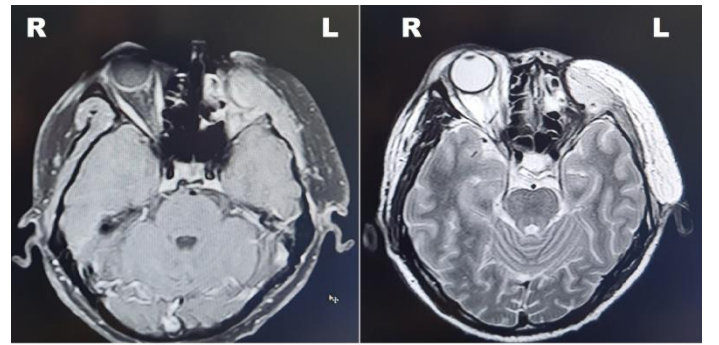


Figure 2: Post treated Magnetic Resonance Imaging (MRI) scan of Face and Neck revealed no residual or recurrent mass seen in post-operative bed. There was myocutaneous free flap in the surgical bed in left frontotemporal scalp extending to zygomatic region and covering entire left orbit (which was exenterated) with post RT changes.



Figure 3: Post Treated photograph of patient.

After that, the patient underwent surgery. Wide Local excision (WLE) of tumor was done including orbital exenteration, left zygoma excision, left level I-IV neck dissection, left superficial parotidectomy (pT4aN₀) on 11th September 2022. Histopathology examination of operated tissue was epithelioid sarcoma with tumor cells positive for CK8/18, CD53, FLI-1, CD99, CD10, SMA, P53, CD68 in scattered cells, INI-1 Lost, ki67 30%, focal positive for CK AE1/AE3, Negative for S100, CK5/6, ALK, CD31, D2-40, Bcl2, h-caldesmon & SOX 10. Grade III, left temporal region, 6 X 6 X 2.5 cm, DOI 1.8 cm, LVSI was not identified. PNI present, margins were uninvolved, 0.1 cm from closest margin, Zygomatic bone was involved by tumor and eyebrow with eyelid was also involved with tumor however epidermis free from tumor (LN0/40, pT_{4a}N₀). Adjuvant Radiation Therapy (RT) 6000 cGy radiation dose in 30 fractions (#) which was 200 cGy per fraction (#) were given during 22nd September to 10th November 2022. Patient was asked to follow up after 6 weeks after completion of RT. After 6 weeks of radiation therapy (RT), the patient undergone for MRI of Head and Neck. MRI examination revealed no residual or recurrent mass seen in the post operative bed. There was Myocutaneous free flap in the surgical bed in left

frontotemporal scalp extending to zygomatic region and covering entire left orbit (which was exenterated) with post RT changes. There was B/L maxillary left ethmoid sinusitis. Computed Tomography (CT) scan Chest was also done to examine lung metastases and revealed that there were no lungs metastases.

Discussion

There are many assumption concerning the derivation and tumorigenesis of epithelioid sarcoma (ES). It has been projected that ES is a mesenchymal carcinoma originated from prehistoric mesenchymal or myofibroblast cells.¹ There is another hypothesis squabble that ES may be originate from naive synovial cells and correspond to a modification of synovial sarcoma.¹⁹ Both subtypes (proximal and distal) of ES influence patients of a broad age variety which is also summarized in table 1. ES is in attendance obvious malignant histologic features, including a high mitotic index, necrosis, and invasiveness. The age at the diagnosis of ES which is originated at scalp ranges from 1 year to 80-years, and the average age are 28 years. The propensity for local recurrence and distal metastasis is higher in ES of the scalp and is usually correlated with bone invasion, indicating its distinction from ES of other locations. ES occurring in the head and neck are supposed to be differentiated from other epithelioid tumors by INI stains.²⁰

The clinical, pathological and other details of previous published case studies and our present case study are summarized (Table 1). In our study, we also summarized literatures review. A total of 15 case studies are included in this literature review including our study. The male female proportion is approximately 2.7:1. The 15 cases studies of scalp ES including our case study preponderated by epithelioid tumor cell morphology. Prognosis of scalp ES is dependent upon various factors

including TNM staging of tumor i.e. tumor size, bony erosion, adjacent organ involvement, lymph nodes involvements and distance metastases. The prognosis of scalp ES is correlated with a poor prognosis and all relapsed cases exhibited bone erosion. Necrosis exists in up to 70% of the cases and is usually associated with central degeneration. Most cases exhibit a dominated epithelioid morphology in the dermis and subcutaneous tissue with ulcer and lymphatic vascular invasion. Granuloma-like features, calcification and bone formation occasionally occur. Approximately, 60 % of cases were recovered from disease including our case study in literature review of all 15 cases. This may be due to external appearance and diagnosed earlier.

Treatment protocol is still not consistent, but radical surgery is the best option for ES scalp. Radiotherapy is given for local invasion of disease. In some cases, chemotherapy is also given but it is not significantly approachable. Many research studies articles have reported on epithelioid sarcoma survival rates through the years. Due to rare occurrences of the disease, the numbers show a discrepancy and significantly. Five-year survival and ten-year survival rate for patients with epithelioid sarcoma are approximately 50-70% and 42-55% respectively.⁵ Our case was a survival case from ES in the scalp.

5	21/F	Supra-auricular	ES; CN, thin fibrosis	Recur in 10 years	Hanna SL/2002 [10]
6	7/M	Scalp	ES; CN, thin fibrosis	FD, 72 months	Hanna SL/2002 [10]
7	32/M	Parietal Scalp	ES; CN		Tocco I/2012 [11]
8	34/F	Lt Temporal Scalp	ES, CN, pseudogranulomatous	FD 18 months	Kim YS/2018 [12]
9	80/M	Scalp	ES; CN, pseudogranulomatous	FD 12 months	Zhang XW/2019 [13]
10	18/M	Occipital Region	ES, hemorrhage; necrosis		Gurwale SG/2019 [14]
11	16/M	Frontoparietal Scalp	ES;	Lung mets 6 months	Bhanu Prakash/2020 [15]
12	67/M	Rt Parietal Scalp	High grade ES	FD 8 months	Zhang Y/2021 [16]
13	47/M	Maxillary/Eyelid	ES; Orbital invasion	FD	Han MM/2023 [17]
14	17/F	Rt Frontal Scalp	ES;	FD	Almasarei HM/2023 [18]
15	36/M	Lt Frontotemporal Scalp	ES; Eyelid and Orbit involvement	FD 6 months	Present Study
Epithelioid sarcoma (ES), Central necrosis (CN), Free of Diseases (FD), Recurrence (Recur)					

Conclusion

To our knowledge, this report describes a unique clinical case and contains the largest descriptive analysis of reported cases of primary non-metastatic scalp ES. We presented the initial clinical findings and treatment course for a male patient diagnosed with ES of the left Fronto parietal scalp, stage pT2 N0 M0. Based on the previously reported clinical outcomes for patients with primary scalp ES. In prior reported clinical outcomes for scalp ES cases, we feel it is reasonable to consider surgical resection followed by postoperative external beam radiotherapy for localized scalp ES. Future research

S N	Age/ Sex	Position	Histopathology	Prognosis	References
1	1/M	Forehead		FD 24 months	Kodet R/1994 [6]
2	7/M	Scalp	ES; CN	FD 15 months	Gross E/1996 [7]
3	47/M	Scalp	ES; High grade		Skoog L/1999 [8]
4	30/F	Parietooccipital scalp	ES;	Recur in 3 rd year	C Suwantemee /1999 [9]

is still needed for this rare malignancy and clinical presentation.

References

- Enzinger FM. Epithelioid sarcoma. A sarcoma simulating a granuloma or a carcinoma. *Cancer*. 1970 Nov;26(5):1029-41. doi: 10.1002/1097-0142(197011)26:5<1029::aid-cnrcr2820260510>3.0.co;2-r. PMID: 5476785.
- Elsamna ST, Amer K, Elkattawy O, Beebe KS. Epithelioid sarcoma: half a century later. *Acta Oncol*. 2020 Jan;59(1):48-54. doi: 10.1080/0284186X.2019.1659514. Epub 2019 Sep 3. PMID: 31478778.
- Steinberg BD, Gelberman RH, Mankin HJ, Rosenberg AE. Epithelioid sarcoma in the upper extremity. *J Bone Joint Surg Am*. 1992 Jan;74(1):28-35. PMID: 1734011.
- Jawad MU, Extein J, Min ES, Scully SP. Prognostic factors for survival in patients with epithelioid sarcoma: 441 cases from the SEER database. *Clin Orthop Relat Res*. 2009 Nov;467(11):2939-48. doi: 10.1007/s11999-009-0749-2. Epub 2009 Feb 18. PMID: 19224301; PMCID: PMC2758965.
- Bos GD, Pritchard DJ, Reiman HM, Dobyns JH, Ilstrup DM, Landon GC. Epithelioid sarcoma. An analysis of fifty-one cases. *J Bone Joint Surg Am*. 1988 Jul;70(6):862-70. PMID: 3392084.
- Kodet R, Smelhaus V, Newton WA Jr, Hamoudi AB, Qualman SJ, Singley C, Jacobs DL. Epithelioid sarcoma in childhood: An immunohistochemical, electron microscopic, and clinicopathologic study of 11 cases under 15 years of age and review of the literature. *Pediatr Pathol*. 1994 May-Jun;14(3):433-51. doi: 10.3109/15513819409024274. PMID: 7520581.
- Gross E, Rao BN, Pappo A, Bowman L, Shearer P, Kaste S, Greenwald C, Michalkiewicz E, Pratt C. Epithelioid sarcoma in children. *J Pediatr Surg*. 1996 Dec;31(12):1663-5. doi: 10.1016/s0022-3468(96)90043-3. PMID: 8986982.
- Skoog L, Pereira ST, Tani E. Fine-needle aspiration cytology and immunocytochemistry of soft-tissue tumors and osteo/chondrosarcomas of the head and neck. *Diagn Cytopathol*. 1999 Mar;20(3):131-6. doi: 10.1002/(sici)1097-0339(199903)20:3<131::aid-dc4>3.0.co;2-t. PMID: 10086236.
- Suwantemee C. Primary epithelioid sarcoma of the scalp. *Plast Reconstr Surg*. 1999 Sep;104(3):785-8. doi: 10.1097/00006534-199909030-00026. PMID: 10456531.
- Hanna SL, Kaste S, Jenkins JJ, Hewan-Lowe K, Spence JV, Gupta M, Monson D, Fletcher BD. Epithelioid sarcoma: clinical, MR imaging and pathologic findings. *Skeletal Radiol*. 2002 Jul;31(7):400-12. doi: 10.1007/s00256-002-0509-9. Epub 2002 May 8. PMID: 12107573.
- Tocco I, Bassetto F, Vindigni V. Primary epithelioid sarcoma of the scalp complicated by humoral hypercalcemia of malignancy. *Eplasty*. 2012;12:ic18. Epub 2012 Dec 10. PMID: 23359842; PMCID: PMC3552616.
- Kim YS, Kwak HS, Chung GH, Kim YN, Hwang S. Epithelioid sarcoma arising from the temporal space: A case report. *Medicine (Baltimore)*. 2018 Sep;97(38):e12529. doi: 10.1097/MD.00000000000012529. PMID: 30235775; PMCID: PMC6160058.
- Zhang XW, Deng YJ, Zhou L, Deng H. Epithelioid sarcoma of the scalp: a case report and literature review. *Int J Clin Exp Pathol*. 2019 Oct 1;12(10):3908-3914. PMID: 31933781; PMCID: PMC6949740.
- Gurwale SG, Gore CR, Kaur S, Deshpande AH. Primary epithelioid sarcoma of scalp. *Asian J Oncol* 2017;3:71-3.
- Bhanu Prakash, Abhishek Budharapu, Umanath K. Nayak, Rahul Buggavetti, Vijay Jain and Ashwin Shastri 2020. Sarcomas of the scalp: a case series. *International Journal of Oral and Facial Surgery*. 2, 1 (Oct. 2020), 04-09. DOI: <https://doi.org/10.33974/ijofs.v2i1.211>.
- Zhang Y, Mirpuri TM, Ho CL. Primary Epithelioid Sarcoma Manifesting as a Fungating Scalp Mass - Imaging Features and Treatment Options. A Case Report and Literature Review. *J Radiol Case Rep*. 2021 Nov 1;15(11):1-9. doi: 10.3941/jrcr.v15i11.4326. PMID: 35516973; PMCID: PMC9063832.
- Han MM, Dermarkarian CR, Camacho MB, Estopinal MDV, Tao JP. Metastatic Epithelioid Sarcoma of the Eyelid in a 47-Year-Old Man: A Case Report. *Ophthalmic Plast Reconstr Surg*. 2023 Jul-Aug 01;39(4):e112-e115. doi: 10.1097/IOP.0000000000002370. Epub 2023 Mar 9. PMID: 36893069.
- Almasarei et al. Epithelioid Sarcoma of the Skull: A Differential Diagnostic Approach, *Journal of the Bahrain Medical Society* 2023; 35 (1): 68-74. https://doi.org/10.26715/jbms.35_1_9
- Patchefsky AS, Soriano R, Kostianovsky M. Epithelioid sarcoma: ultrastructural similarity to nodular synovitis. *Cancer*. 1977 Jan;39(1):143-52. doi: 10.1002/1097-0142(197701)39:1<143::aid-cnrcr2820390125>3.0.co;2-3. PMID: 188535.
- Frank R, Sadri N, Bhatti T, Biegel JA, Livolsi VA, Zhang PJ. Proximal-type Epithelioid Sarcoma of the Head and Neck (HN): A Study with Immunohistochemical and

Molecular Analysis of SMARCB1. J ClinExpOncol.
2013;2(2):1000106. doi: 10.4172/2324-9110.1000106.

PMID: 24308011; PMCID: PMC3846256.