The diagnosis of Paget's disease is confirmed by the presence of Paget cells, which are round with clear cytoplasm and large pleomorphic nuclei [1]. Immunohistochemical confirmatory antigens are carcinoembryonic antigen and low molecular weight CK such as CK 7, indicating the glandular origin of Paget's disease [1]. Other antigens that can be used to distinguish an apocrine origin from an eccrine origin are apocrine epithelial antigen, human milk fat globule 1, and GCDFP 15, and these antigens are routinely identifiable in Paget cells [1].

The present patient showed obvious brain metastasis from EMPD with histopathological findings including the above typical EMPD immunohistochemical profile. Additionally, we identified worsening of the patient's skin lesions during a 3 month interval, and histopathological findings included tumor nests in the lymphatic channel, which were also metastatic lesions. Extension of the regions of tracer uptake to multiple lymph nodes in the trunk may have been an early sign of brain metastasis from EMPD in this patient. Co-existence of another systemic malignancy was excluded based on the absence of uptake in the visceral organs on serial PET images taken over the 3 month interval.

No definitive guidelines are available to manage brain metastasis from EMPD. However, radiation therapy is an established treatment for EMPD, [7] and the indications for large or inoperable tumors and patients with postoperative recurrence [8]. Takahagi et al. reported a patient with EMPD with suspected brain metastasis on MRI [3]. They managed the patient with brain radiation and systemic chemotherapy with 5-fluorouracil and cisplatin. Despite an absence of follow-up imaging, they reported that radiation therapy prevented the brain metastasis from growing rapidly [3]. In our case, although the patient died of acute respiratory failure, we identified an effect of radiation therapy on the brain metastasis from EMPD on the MRI performed before and after radiation therapy [7].

In conclusion, we report to our knowledge the first patient with histopathologically proven brain metastasis from EMPD. The brain metastasis was rapid and aggressive, and simultaneous with worsening of skin lesions during a 3 month interval. Additionally, we radiologically identified for the first time an effect of radiation therapy for an EMPD brain metastasis.

Conflict of interest/disclosure

The authors declare that they have no financial or other conflicts of interest in relation to this research and its publication.

Appendix A. Supplementary material

Supplementary data associated with this article can be found, in the online version, at http://dx.doi.org/10.1016/j.jocn.2013.05.027.

References

[1] Wagner G, Sachse MM. Extramammary Paget disease – clinical appearance, pathogenesis, management. J Dtsch Dermatol Ges 2011;9:448–54.
[2] Kim JC, Kim HC, Jeong CS, et al. Extramammary Paget's disease with aggressive behavior: a report of two cases. J Korean Med Sci 1999;14:223–6.
[3] Takahagi S, Noda H, Kamegashira A, et al. Metastatic extramammary Paget's disease treated with paclitaxel and trastuzumab combination chemotherapy. J Dermatol 2009;36:457–61.
[4] Okuno H, Nishio Y, Takeuchi H, et al. Genital Paget's disease with general metastasis: a case report. Hinyokika Kiyo 1988;34:714–7.
[5] Yugueros P, Keeney GL, Bite U. Paget's disease of the groin: report of seven cases. Plast Reconstr Surg 1997;100:336–9.
[6] Cappuccini F, Tewari K, Rogers LW, et al. Extramammary Paget's disease of the vulva: metastases to the bone marrow in the absence of an underlying adenocarcinoma-case report and literature review. Gynecol Oncol 1997;66:146–50.
[7] Brown RS, Lankester KJ, McCormack M, et al. Radiotherapy for perianal Paget's disease. Clin Oncol 2002;14:272–84.
[8] Luk NM, Yu KH, Yeung WK, et al. Extrammamary Paget's disease: outcome of radiotherapy with curative intend. Clin Exp Dermatol 2003;28:360–3.

http://dx.doi.org/10.1016/j.jocn.2013.05.021

Giant calvarial desmoplastic fibroblastoma

Ravi Dadlani [a], Nandita Ghosal [b,*], Alangar S. Hegde [a], Kanchan Gupta [c]

[a] Department of Neurosurgery, Sri Satya Sai Institute of Higher Medical Sciences, Bangalore, India
[b] Department of Pathology, Sri Satya Sai Institute of Higher Medical Sciences, EPIP Area, Whitefield, Bangalore 560 066, India
[c] Department of Radiology, Sri Satya Sai Institute of Higher Medical Sciences, Bangalore, India

ARTICLE INFO

Article history:
Received 19 February 2013
Accepted 15 May 2013

Keywords:
Calvarium
Childhood
Desmoplastic fibroblastoma
Female
Skull

ABSTRACT

We describe to our knowledge the first patient with giant desmoplastic fibroblastoma of the calvarium in a 20-year-old woman whose tumor first appeared at the age of 5 years. We also discuss the histopathological differential diagnosis, management dilemmas and complications of desmoplastic fibroblastoma.

1. Introduction

Desmoplastic fibroblastoma (DF) was first described by Evans in 1995, and was renamed as collagenous fibroma by Nielsen in 1996 [1]. We report a young woman with a giant calvarial DF with involvement of the overlying scalp.

* Corresponding author. Tel.: +91 080 2841 1500x316; fax: +91 080 2841 1502.
E-mail address: nandita.g@rediffmail.com (N. Ghosal).

2. Case report

A 20-year-old woman presented with a history of progressive swelling on the anterior scalp measuring 15 × 12 × 12 cm, first noticed at the age of 5 years (Fig. 1). Plain radiographs of the skull revealed a large sclerotic lesion involving the frontal bone with presence of "sun ray" spicules with significant soft tissue component (Fig. 2a, b). A CT scan demonstrated spiculations arising from the outer table, which were further delineated in the three-dimensional reconstruction (Fig. 2d, e). MRI of the skull revealed a giant heterogeneous lesion measuring 18 × 14 × 17.5 cm involving both frontal bones and bilateral superior orbital ridges, which was markedly hypointense on both T1-weighted and T2-weighted sequences, with a soft tissue component that was isointense on T1-weighted and heterointense on T2-weighted images (Fig. 2c, f). Multiple hypointense bony "sun ray" spicules were noted to be extending into the soft tissue component.

The patient underwent surgical excision of lesion along with reconstructive surgery. Only subtotal excision of the lesion was possible in view of the increased vascularity (Fig. 1e). Intra-operatively there was diffuse oozing in addition to several large supplying arteries from the middle meningeal and the external carotid artery circulation. Intra-operatively the lesion was seen to arise from the calvarium because the scalp was completely free from infiltration and could be easily separated from the skull as in routine surgery. The lesion was diffusely infiltrating the bone and had no capsule (Fig. 3a). On gross examination the lesion consisted of two main parts: the soft outer and hard bony deeper part from which the bony spicules were arising (Fig. 3b). It was gritty on cut section and appeared grey-white with a firm consistency. Tissue sent for histopathological analysis was routinely processed with decalcification (10% hydrochloric acid) of the bony component. Five micron thick sections were cut from paraffin embedded tissue and stained with hematoxylin and eosin. On light microscopy a paucicellular lesion admixed with an eosinophilic extracellular matrix was seen. Cells were spindle and stellate shaped with delicate, vesicular nuclei dispersed in abundant glassy keloid like collagen (Fig. 4A, B) highlighted by Masson's trichrome stain as prominent blue colored stroma (Fig. 4C). Focal osseous metaplasia with few prominent dilated vessels with perivascular hyalinization was seen. The lesion was infiltrating in between the bony trabecu-

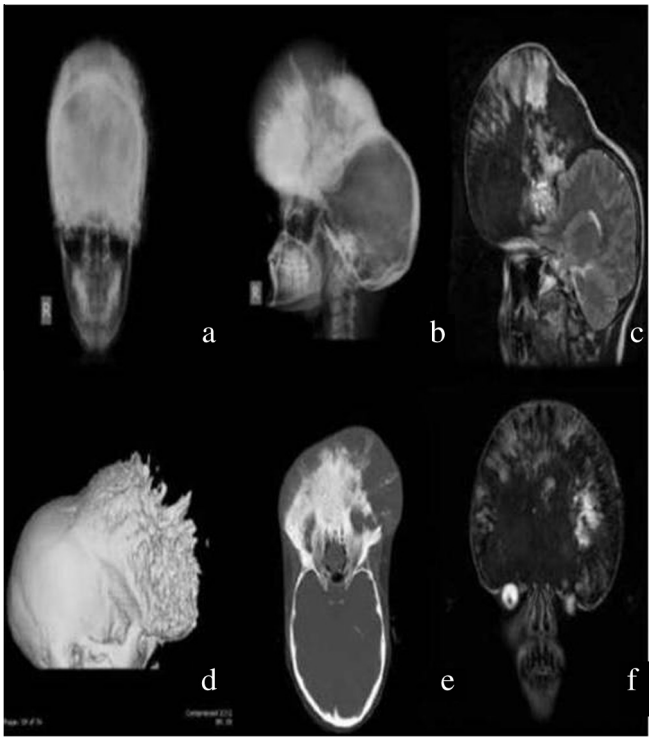

Fig. 2. Plain (a) anteroposterior and (b) lateral radiographs showing the extent of the lesion and sclerotic spicules. T2-weighted (c) sagittal and (f) coronal MRI of the brain showing a heterogeneous lesion involving the bilateral superior orbital ridges. CT (d) three dimensional surface shaded reconstruction and (e) axial scans with bone windows showing spiculations arising from the outer table.

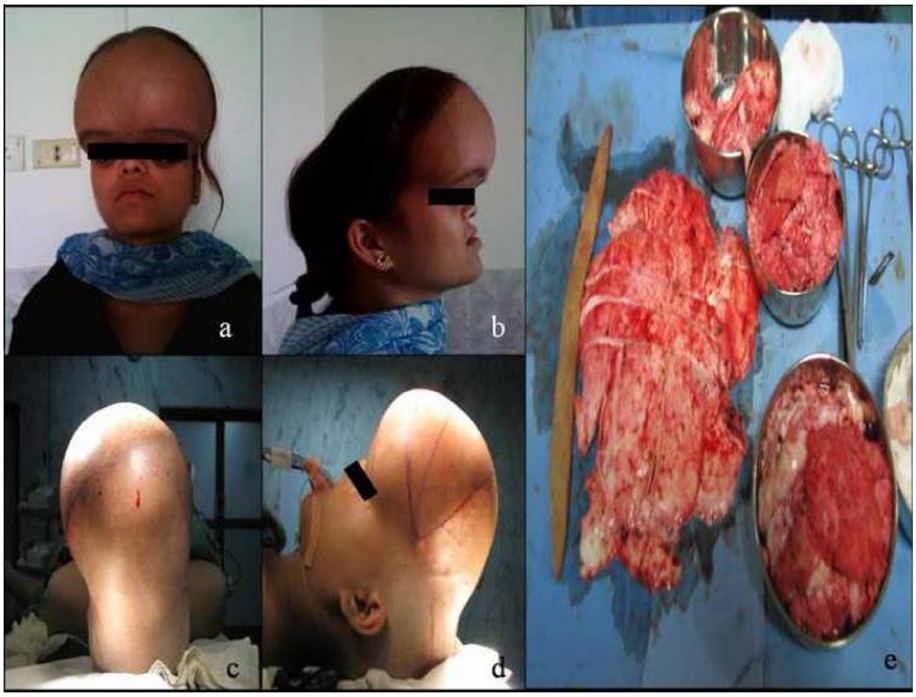

Fig. 1. Patient photographs showing (a, b) the lesion involving supra-orbital ridges and displacing the eyebrows superiorly. Intra-operative photographs showing (c, d) delineating the bicoronal incisions with the redundant skin that was excised, and (e) excised tumor tissue. (This figure is available in colour at www.sciencedirect.com)