Journal of the Egyptian National Cancer Institute 29 (2017) 167–170

Contents lists available at ScienceDirect

Journal of the Egyptian National Cancer Institute

journal homepage: www.sciencedirect.com

Case report

Alveolar soft part sarcoma of orbit: A rare diagnosis

Pritee B. Chaudhari [a], Sushmita Pathy [a,*], Suryanarayana S.V. Deo [b], Bhavna Chawla [c], Asit Ranjan Mridha [d]

[a] Department of Radiation Oncology, Dr. B.R. Ambedkar Institute Rotary Cancer Hospital, All India Institute of Medical Sciences, Ansari Nagar, New Delhi 110029, India
[b] Department of Surgical Oncology, Dr. B.R. Ambedkar Institute Rotary Cancer Hospital, All India Institute of Medical Sciences, Ansari Nagar, New Delhi 110029, India
[c] Department of Ophthalmology, Dr. Rajendra Prasad Ophthalmic Centre, All India Institute of Medical Sciences, Ansari Nagar, New Delhi 110029, India
[d] Department of Pathology, All India Institute of Medical Sciences, Ansari Nagar, New Delhi 110029, India

ARTICLE INFO

Article history:
Received 8 June 2017
Received in revised form 19 July 2017
Accepted 22 July 2017
Available online 23 August 2017

Keywords:
Alveolar soft part sarcoma
Orbit
Adjuvant
Radiotherapy

ABSTRACT

Objective: Alveolar soft part sarcoma (ASPS) is an aggressive, rare tumour with unique morphological and histopathological features.
Methods: We report a rare case of orbital ASPS and its management in a young male who presented with painless proptosis and progressive loss of vision.
Result: Twenty-two year male presented with a history of gradually increasing proptosis with loss of vision since 12 months. He underwent radical re-excision of mass with right orbital exenteration and reconstruction using temporalis muscle flap. Adjuvant radiotherapy to a dose of 64 Gy in 32 fractions over 6.5 weeks was planned in view of positive surgical margins. Patient is free of disease and currently under follow up in multidisciplinary clinic.
Conclusion: Function preserving surgery remains the standard treatment approach in localised disease however the complex anatomy and locally aggressive nature makes it difficult to achieve clear surgical margin. Adjuvant radiotherapy has shown to improve local control in patients with positive surgical margins.
© 2017 National Cancer Institute, Cairo University. Production and hosting by Elsevier B.V. This is an open access article under the CC BY-NC-ND license (http://creativecommons.org/licenses/by-nc-nd/4.0/).

Introduction

Alveolar soft part sarcoma (ASPS) is a rare tumour with unique morphological and histopathological features. It accounts for less than 1% of soft tissue sarcomas and affects commonly young individuals during second and third decade of life with a female preponderance [1]. ASPS commonly occur in trunk and extremities [1,2]. Other sites include retroperitoneum, pelvis, head, neck, mediastinum, bladder, gastrointestinal tract [2,3]. The tumour has an indolent course often delaying diagnosis and treatment. Orbital ASPS is a rare entity with few isolated published reports [4,5]. Here in we report a case of orbital ASPS and its management in a young male who presented with painless proptosis and progressive loss of vision.

Case presentation

A 22-year-old male presented with 12 months history of proptosis and progressive loss of vision in right eye. The symptoms worsened in last 4 months for which he sought ophthalmic consultation. After an initial biopsy he was referred to Radiation Oncology clinic for adjuvant therapy. Physical examination revealed his performance status 1 (Eastern Cooperative Oncology Group) Ocular examination revealed, absence of Perception of light (PL) and projection of rays (PR) in the right eye. Proptosis was severe (30 mm) unilateral, and eccentric (globe pushed downward and outward), The mass was soft, 3 × 3 cm, compressible. Orbital margin was intact. No bruit was present on auscultation. There was no regional lymphadenopathy. Ocular motility was grossly restricted on elevation and adduction. Pupillary reflex was absent. Examination of left eye did not reveal any abnormalities.

Investigations and treatment

Contrast enhanced computed tomography of orbit, face and neck showed 5 × 4.7 × 3.7 cm heterogeneously enhancing exophytic, extraconal mass in superior part of right orbit involving the superior rectus, oblique muscles and optic nerve. Thinning

Peer review under responsibility of The National Cancer Institute, Cairo University.
* Corresponding author.
E-mail addresses: drpritee184@gmail.com (P.B. Chaudhari), drspathy@gmail.com (S. Pathy), svsdeo@yahoo.co.in (S.S.V. Deo), bhavna2424@hotmail.com (B. Chawla), asit_aiims@yahoo.co.in (A.R. Mridha).

http://dx.doi.org/10.1016/j.jnci.2017.07.001
1110-0362/© 2017 National Cancer Institute, Cairo University. Production and hosting by Elsevier B.V.
This is an open access article under the CC BY-NC-ND license (http://creativecommons.org/licenses/by-nc-nd/4.0/).

and remodelling of medial wall of orbit was observed. There was no intracranial or sinonasal extension. Mass involved the bellies of both lateral and medial rectus (Fig. 1). Rest of the metastatic work-up revealed no abnormality. Patient was staged as T1N0M0; stage IA according to AJCC 2010 staging.

As the initial excision appeared inadequate, re-excision of the mass with orbital exenteration and reconstruction using temporalis muscle flap through a lateral orbitotomy approach was performed after obtaining informed consent.

Operative details included extraocular vascular lesion in superomedial aspect of right orbit pushing globe inferiorly and outwards and infiltrating the outer table of roof of orbit. Pressure destruction of medial wall of bony orbit was present. Exenteration involved removal of the contents of the eye socket including muscles, lacrimal gland, optic nerve and bones comprising of medial wall of orbit. Reconstruction was performed with temporalis muscle. Postoperative gross specimen showed a grey white fleshy tumour of size $5 \times 4.6 \times 4$ compressing the eyeball in extraocular spaces. Histopathological examination revealed tumour arranged in solid nest separated by thin fibrovascular septae and large uniform epithelioid cells with abundant eosinophilic granular cytoplasm and a centrally placed nucleus with prominent nucleolus. The posterior resected margin was involved by tumour with close superior and medial margins**.** The lacrimal bone also showed positive tumour infiltrates. There was no intraocular involvement. The tumour cells were immunonegative to pan-cytokeratin (1:250) (Bio SB, US), HMB-45 (1:100) (Thermo Fisher Scientific Inc, US), S-100 (1:800) (Dako, US), desmin (1:100) (Thermo Fisher Scientific Inc, US) and myogenin (1:50) (Thermo Fisher Scientific Inc, US). Vascular markers like CD 31 and CD 34 were negative. Final diagnosis of alveolar soft part sarcoma was made on the basis of histological and immunohistochemistry findings (Fig. 2). Adjuvant radiotherapy was planned in view of positive and close surgical margins. Linac based three-dimensional conformal technique (Elekta Medical Systems Crawley, UK) was delivered to the right orbit to a dose of 64 Gy in 32 fractions over six and half weeks with right medial oblique and right lateral oblique fields with 6 MV photons. The entire orbit was contoured to form the clinical target volume and the planning target volume was formed with a (0.5 cm) margin to encompass set up error. Radiation dose to surrounding organs and normal organs at risk were maintained within acceptable limits.

Outcome and follow up

The treatment was well tolerated with acceptable morbidity. At six months following treatment patient is currently disease free and is under regular follow-up for local and distant failure.

Discussion

ASPS is a rare tumour accounting for less than 1% of all soft tissue sarcoma with an uncertain histology. The terminology, ASPS was first coined by Christopherson et al. [6]. Prior reports include case reports and series spanned over a decade including mostly extremities and trunk. Orbital ASPS is extremely rare with a peak incidence between 15 and 35 years [7]. The case reported here is in the second decade. Orbital involvement presents with slow growing mass resulting in proptosis, globe displacement, ptosis with loss of vision [8,9]. Similar symptoms and clinical course was observed in the current report.

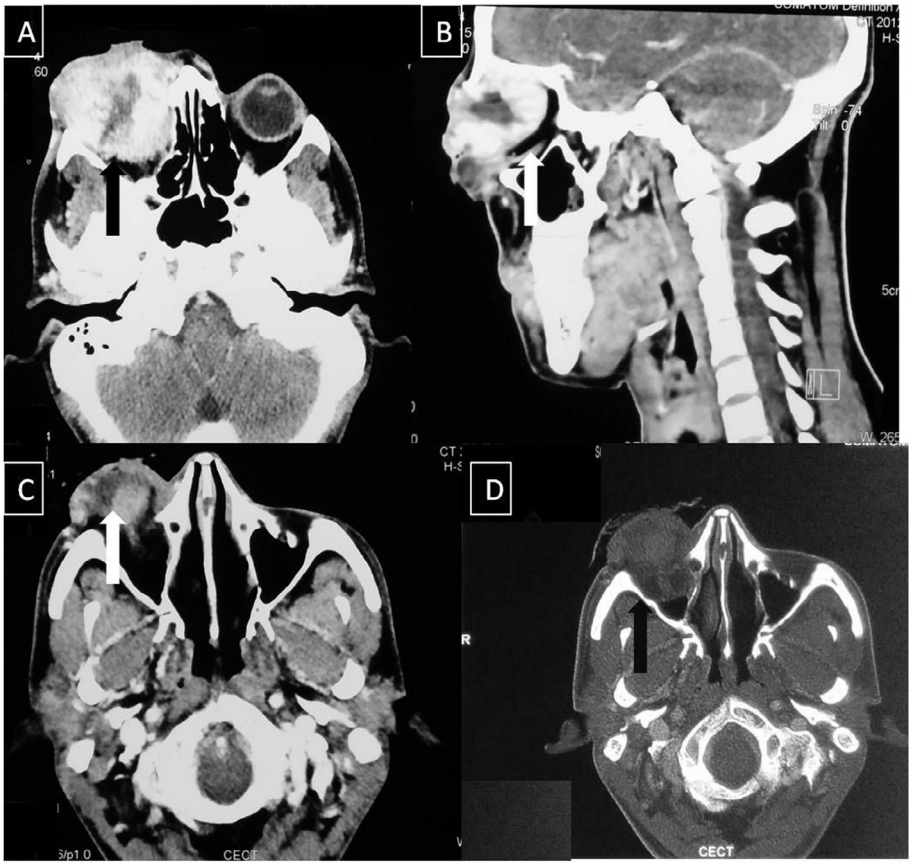

Fig. 1. A = Enhancing lobulated extraconal mass in superior part of right orbit with central areas of necrosis. B = Mass is displacing the globe inferiorly and anteriorly without involving the optic nerve. No intracranial extension seen. C = Mass is extending into right globe. D = No bony destruction and no sinonasal extension present.

Download English Version:

https://daneshyari.com/en/article/8787482

Download Persian Version:

https://daneshyari.com/article/8787482

Daneshyari.com