ISSN: 2634 - 680X

Journal of Clinical Case Studies Reviews & Reports

SCIENTIFIC Research and Community

Case Report

Open Access

Primitive Neuroectodermal Tumor of Axilla in an Adult: A Rare Entity

Shetty Shravan S[1]*, Shah Vidhi[2], Eshpuniyani Prriya[3] and Pathan Sameer[4]

[1-3]Department of Surgical Oncology, Asian Cancer Institute, Mumbai, India

[4]Department of Pathology, Asian Cancer Institute, Mumbai, India

SUMMARY

Primitive neuroectodermal tumors (PNETs) are poorly differentiated small round cell neoplasms which mainly affect children and are not commonly seen in adults. Superficial primitive neuroectodermal tumors are rare and have a favourable prognosis compared to conventional deep seated tumors. We report a case of a 62 year old gentleman with a primitive neuroectodermal tumor arising from the subcutaneous tissue of the right axilla. He was treated with multimodal treatment including surgery and radiotherapy. He is alive and disease free at 2 year follow up.

***Corresponding author**

Shetty Shravan, Department of Surgical Oncology, Asian Cancer Institute, Mumbai, India. E-mail: drshravanshetty@gmail.com

Received: February 25, 2021; **Accepted:** March 02, 2021; **Published:** March 07, 2021

Keywords: Ewing's Sarcoma, PNET, Subcutaneous, Surgery, Prognosis

Introduction

Ewing's Sarcoma was first described by James Ewing in 1921 and it was initially distinguished from PNET as a different entity [1]. However it contain the same reciprocal translocation between chromosomes 11 and 22 (11; 22). Both entities are now included in a broader category called Ewings family of tumors (EFST). EFST includes Ewing's Sarcoma of bone, Extraosseous Ewing's Sarcoma (EES), PNET and Askin tumors. Although majority of extraskeletal ES/PNET are located in the deep soft tissues, there are a few published cases of primary cutaneous ES/PNETs which are slow growing and have a better prognosis with reported survival rates of 91% in 10 years [2-4].

Clinically primary cutaneous and subcutaneous Ewing's sarcoma (scEWS) afflicts children and young adults. scEWS most commonly presents as 2 to 3 cm (range 0.5 – 12cm) superficial mass on the trunk or lower /upper extremities of young females [5]. EES is usually established by typical features on histologic analysis, histochemistry, immunohistochemistry and electron microscopy [6,7]. Differential diagnosis include other small round blue cell tumors and other members of the Ewing's family of tumors [8].

Treatment is usually multimodality including surgery, radiotherapy and chemotherapy. The gold standard for localised disease remains surgery and may need adjuvant radiotherapy and chemotherapy.

Case Report

A 67 year old gentleman presented with a painless swelling in the right armpit since 6 months. On examination, he had an approximately 8x8 cm firm palpable lump in right axilla with a biopsy scar over it. Rest of physical examination was normal. An incision biopsy of the lump done elsewhere revealed Ewing's Sarcoma showing prominent epithelial differentiation. On FISH test, EWSR1 gene rearrangement was detected.MRI showed a solid right axillary lesion with a small cystic component over the surrounding tissue. There was no invasion of surrounding muscles, brachial plexus or axillary vessels. PET CT showed a right axillary lesion with SUV Max 11.05 with no distant metastases (Figure 1 & 2). He was planned for surgery followed by adjuvant treatment at multidisciplinary tumour board meeting. He underwent wide excision of the axillary lump along with scar excision (Figure 3). Final histopathology report showed a 9.4 cm, large malignant small round cell tumour with areas of necrosis. Surgical margin of resection were free. Mitotic activity is high with average of 4-5/hpf. All lymph nodes were negative. Immunohistochemistry studies showed CD 99+, FL-1+ & VIMENTIN+ which confirmed the diagnosis of primitive neuroectodermal tumor (pT2aN0M0). Post surgery, the patient received adjuvant radiotherapy. He is disease free at 2 year follow up.

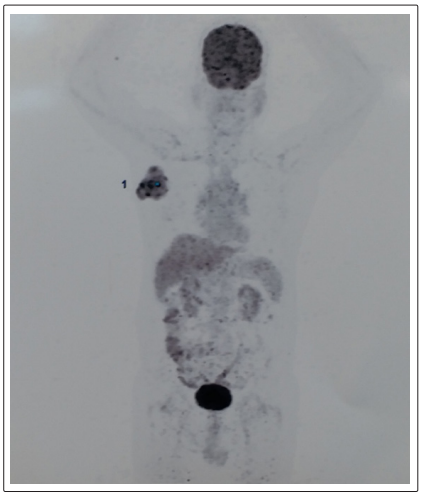

Figure 1

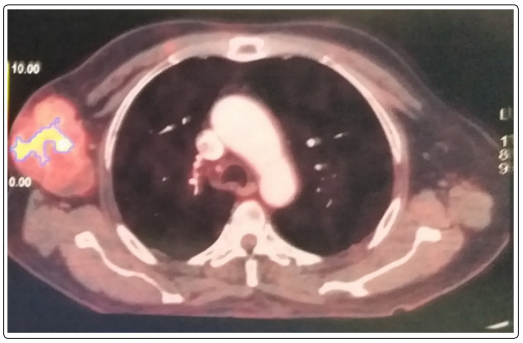

Figure 2

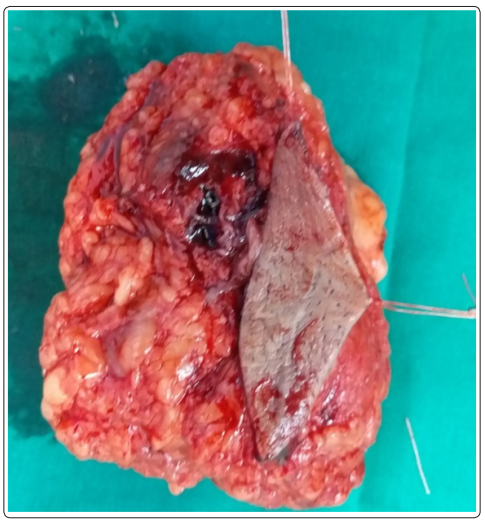

Figure 3

Discussion

EWS of the bone, EES and sc EWS share similar molecular and cytogentic characteristics, including strong membranous expression of CD99 (MIC2) and the presence of a EWSR translocation involving chromosome 22 [5]. Both these characteristics were seen in this case. Frequently affected extraskeletal sites include paravertebral spaces, lower extremities, head & neck and pelvis [9]. Other uncommon locations of EES include retroperitoneum, omentum, orbit, skin and chest wall [10]. There have been reports of skin and subcutaneous PNETs mainly in children, adolescents and young adults however to the best of our knowledge axillary subcutaneous PNET in adults has not been reported [11].

As per NCCN guidelines, the same treatment algorithm for all members of Ewings family of tumors can be used. The treatment recommended is local treatment (surgery and / or radiotherapy) plus chemotherapy. Rud et al proposed that surgery may have a more significant role in EES compared to skeletal ES and that total resection predicts good survival [12]. Most cases reported in literature are treated as the rest of the ESFT, with surgery when possible and a combination with chemotherapy or radiotherapy [13,14].

Our patient underwent complete local excision followed by adjuvant radiotherapy. scEWS has a 4 to 5 year overall survival of 91-94% and a 5 year event free survival of 88.5% [4,5]. Good prognosis has also been attributed to early detection in sc EWS with superficial scEWS having best prognosis. Our patient is a case of localised pT2aN0Mx scEWS and is disease free at 24 month follow up.

References

1. Ewing J (1921) Diffuse endothelioma of bone. Proc n Y Pathol Soc 21: 17.
2. Cash T, Mcllvaine E, Segal MR, Stephen L. Lessnick,3 Elizabeth R. Lawlor, et al. (2016) Comparison of clinical features and outcomes in patients with extraskeletal versus skeletal localized Ewing sarcoma: A report from the Children´s Oncology Group. Pediatr Blood Cancer 63: 1771-1779.
3. Jaime de Oliveira Filho, Ana Rita ferrante Mitidieri de Oliveira, Natalie Haddad, Ana Carolina franco Tebet, Kassila Nasser (2014) Primary Cutaneous Ewing sarcoma-Case report. Ann Bras dermatol 89: 501-503.
4. M.Delaplace, C.Lhommet, G. De pinieux, B. Vergier, A. De Muret, et al. (2012) Primary cutaneous Ewing Sarcoma. Br J of Dermatol 166: 721-726.
5. Di Giannatale A, Frezza AM, Le Deley MC, Marec-Bérard P, Benson C, et al. (2015) Primary cutaneous and subcutaneous Ewing sarcoma. Pediatr Blood Cancer 62: 1555-1561.
6. Pinto A, Dickman P, Parham D (2011) Pathobiologic markers of the Ewing Sarcoma family of tumors: state of the art and prediction of behaviour. Sarcoma Article ID 856190.
7. Buch AC, Panicker N, Sarawagi S, Anwekar S, Kharat AT (2010) Fine needle aspiration cytology diagnosis of paravertebral extraosseous Ewing's sarcoma. J Cytol 27:146-148.
8. O Delattre 1, J Zucman, T Melot, X S Garau, J M Zucker, G M Lenoir, et al. (1994) The Ewing family of tumors—a subgroup of small-round-cell tumors defined by specific chimeric transcripts. N Engl J Med 331: 294-299.
9. Cheung CC, Kandel RA, Bell RS, Mathews RE, Ghazarian MD (2001) (2001) Extraskeletal Ewing sarcoma in a in a 77-year-old woman. Arch Pathol Lab Med 125:1358-1360.
10. Geens L, Van Robays J, Geert V, Van der Speeten K (2013) An unusual location of extraosseous Ewing's sarcoma. Case Rep Oncol 6: 293-302.
11. Banerjee SS, Agbamu DA, Eyden BP, Harris M (1997) Clinicopathological characteristics of peripheral primitive neuroectodermal tumour of skin and subcutaneous tissue. Histopathology 31: 355-366.
12. Rud NP, Reiman HM, Pritchard DJ, Frassica FJ, Smithson WA (1989) Extraosseous Ewing's sarcoma. A study of 42 cases. Cancer 64: 1548-1553.
13. R.Cabrera, P.Sánchez, M.A. Rodríguez (2008) Tumor subcutáneo neuroectodérmico primitivo periférico. A propósito de un caso. Actas Dermosifiliogr 99: 555-559.
14. Kourda M, Chatti S, Sfia M, Kraiem W, Ben Brahim E (2005) Primary cutaneous extraskeletal Ewing's sarcoma. Ann de Dermatol et Venereol 132: 986-989.