Primary synovial cell sarcoma of the socket: Case report

Bonsaana GB, Nyenze EM, Gedi SH

Department of Ophthalmology, College of Health Sciences, University of Nairobi, P.O. Box 19676 – 00202, Nairobi, Kenya

Corresponding author: Dr GB Bonsaana**.** Email: bbgily@gmail.com

ABSTRACT

We report the first case of primary synovial cell sarcoma of the socket and the seventh of the globe and orbit in literature. Synovial cell sarcomas are relatively common intermediate to high grade malignant soft tissue tumours often with an initial indolent course and may present as a masquerade syndrome as in this case where a 12 year old female initially presented with a unilateral cataract of the right eye and cataract surgery was done with a posterior capsular intraocular lens implant. The patient was seen about 5 years later with endophthalmitis and anterior staphyloma for which evisceration was done. Seven months later the patient presented yet again with a mass in the right socket. Debulking surgery was done following an incisional biopsy for histopathology which showed a biphasic synovial cell sarcoma. Bcl-2 was positive on immunohistochemistry confirming the diagnosis. The patient was treated with chemotherapy followed by radiotherapy and has been healthy with no evidence of recurrence and is on regular follow up.

Key words: Synovial, Sarcoma, Socket, Masquerade syndrome, Immunohistochemistry

INTRODUCTION

Synovial cell sarcomas are relatively common intermediate to high grade malignant soft tissue tumour often with an initial indolent course. It typically presents in adolescents and young adults (15-40 years of age). There may be a mild (M: F 1.2:1) male predilection. The most common location for these tumours is within the soft tissues adjacent to large joints, such as the knee and popliteal fossa. While these tumours arise near joints, it is rare for them to arise from the joint itself and despite the name; they do not arise from synovial structures, such as joints, tendon sheaths and bursae. Synovial cell sarcomas account for 2.5-10% of all soft tissue sarcomas[2,3]. Its name is derived from the fact that microscopically it resembles normal synovium, however it stains for epithelial markers (epithelial membrane antigen and cytokeratin) unlike synovium. It has also been found in many locations that does not have normal synovium such as the orbit. They have non-specific appearance macroscopically; varying from well to poorly defined heterogeneous masses with frequent areas of haemorrhage and necrosis. These sarcomas are divided histologically into 4 sub types; monophasic fibrous: 50-60%, biphasic: 20-30%, poorly differentiated: 15-25%, monophasic epithelial: very rare[1]. Cytogenetic aberration of the t(X;18) translocation is highly specific, seen in over 90% of cases[2-4].

Synovial cell sarcomas are commonly located at the extremities: 80-95%[4] [lower limb: 60-70%], [upper limb: 15-25%], [from soft tissues: 90-95%), [from joint: 5-10%]; head and neck: 5% [hypopharynx or parapharyngeal space, CNS[1], conjunctiva[2], orbit[5-9]; chest wall and viscera: rare (lung)[3], kidney, prostate, oesophagus, heart, pericardium, pleura)[1]. The differential diagnosis include pleomorphic undifferentiated sarcoma [malignant fibrous histiocytoma (MFH)-fibrosarcoma and liposarcoma], other sarcomas [osteosarcoma and chondrosarcoma] and metastatic carcinoma**.** Treatment involves a combination of surgery and usually adjuvant radiotherapy ± chemotherapy. Radiotherapy is particularly useful in treating tumours where an adequate clear margin cannot be achieved, and ideally radiotherapy is administered pre-operatively. Good prognostic variables include: small size, located in extremity, younger age < 20 years of age, solid homogenous mass, presence of calcification, biphasic histology (controversial). Poor prognostic variables include: large size (> 5cm) - most important factor, located in the trunk or head and neck, older patients, cystic/haemorrhagic components, marked heterogeneity, histology, poorly differentiated histology, rhabdoid cells, extensive tumour necrosis, high nuclear grade, p53 mutations, high mitotic rate (> 10 mitoses/10 high-power field). Overall 5 year survival is between 36-76% with both local recurrence (30-50%) and distant metastases (40%) being common[1].

CASE REPORT

We report a case of a twelve year old female patient, who was referred to the Kenyatta National Hospital (KNH), Nairobi from Garissa Government Hospital (GGH), Kenya, with a right socket mass for further investigations and management in December, 2013. The patient had a history of progressive swelling of the right socket 2 months prior to presentation at GGH. She initially developed poor vision and a white reflex (leukocoria) of the right eye at the age of 8 years and was diagnosed with cataract. Cataract surgery was done with a posterior capsular intraocular lens implant in 2008. Her vision improved. There was no history of fever, anorexia, weight

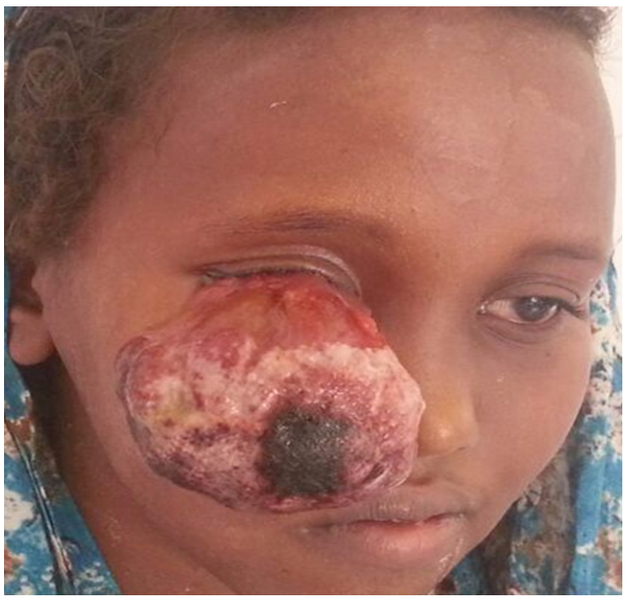
loss, trauma, squint or proptosis. Past medical history was insignificant. No family history of ocular tumour, phthysical eye or blindness. Systemic examination was normal.

In March, 2013, 5 years later, the patient presented to GGH with persistent headache, ocular pain, poor vision and a swelling in the right eye. A diagnosis of endophthalmitis with anterior staphyloma was made and evisceration was done. The patient was well until in October, 2013, 7 months later, when she developed a growth in the right socket which was initially small but rapidly increased in size over a few weeks prompting her to present to GGH again. Systemic examination revealed a right preauricular lymphadenopathy. The patient was referred to KNH for further investigations and management.

In KNH, the patient was in fair general condition with right preauricular lymphadenopathy which had no skin changes or differential warmth, measuring 2mm by 2mm, well defined, firm, non-tender, discrete, mobile and not attached to overlying or underlying structures. There was a mass arising from the right socket free from the eye lids and orbital rim. It measured 18mm by 23mm, reddish in colour, infected and had a whitish discharge, non-tender and soft with a necrotic area overlying it. The left eye vision was 6/6 with normal anterior and posterior segments. Systemic examination was normal.

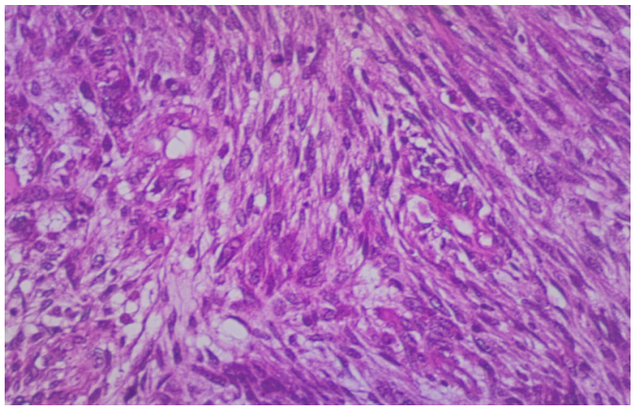
An incisional biopsy of the right socket mass was done for histopathology and immunohistochemistry. The mass dramatically increased in size following the incisional biopsy, measuring 40mm by 50mm, overlying the lower eye lid, right cheek, nose and upper lip (Figure 1). Debulking surgery was done with wide margins.

Figure 1: Patient after incisional biopsy when the lesion developed rapid growth



The histopathology results showed small round blue cells. Biphasic tumour composed of sheets of highly anaplastic spindle cells with irregularly shaped nucleus with high mitotic rate (> 10 mitoses/10 high-power field). The epitheliod component was composed of cellular nests of epitheliod cells that were also seen to form epithelial linings (Figure 2).

Figure 2: Spindle and epitheliod cells of biphasic synovial cell sarcoma of the socket



Immunohistochemistry showed:
Bcl-2 – Positive
Desmin – Negative
EMA - Negative
CD 99 – Negative

A diagnosis of primary orbital synovial cell sarcoma of the socket was made. She was treated with chemotherapy and radiotherapy. She received 12 courses of VAC-Cis (Vincristine, Adriamycin, Cyclophosphamide and Cisplatin). She is on follow up with no signs of recurrence for 2 years now (Figure 3).

Figure 3: Patient after debulking surgery and chemo radiotherapy showing the right socket

DISCUSSION

This patient presented a diagnostic dilemma considering the various pathologies she presented with. It is possible that the cataract and staphyloma may have been different pathologies and unrelated. There have been a few cases of primary synovial cell sarcoma of the orbit[2] and the conjunctiva[2] reported but there has not been a case of the socket following evisceration reported as yet and this might as well be the first case. This is the seventh case of primary synovial cell sarcoma of the globe and orbit reported in literature.

Synovial cell sarcomas are relatively common intermediate to high grade malignant soft tissue tumour often with an initial indolent course as seen in our patient. The name is derived from the fact that microscopically it resembles normal synovium, however it stains for epithelial markers (epithelial membrane antigen and cytokeratin) unlike synovium. It has also been found in many locations that do not have normal synovium such as the socket as in this case. It has non-specific appearance macroscopically, varying from well to poorly defined heterogeneous masses with frequent areas of haemorrhage and necrosis[2-4] as evident in this case report.

CONCLUSIONS

Orbital synovial cell sarcoma is a rare condition with few cases reported in literature. This case had a more complicated past ocular events which might have been unrelated pathologies. Immunohistochemistry was very useful in confirming the diagnosis in our case report. Chemo-radiotherapy is useful in treating incompletely excised primary synovial cell sarcoma.

REFERENCES

1. Weerakkody Y, Gaillard F. Synovial sarcoma. *Radiopaedia* [Internet]. Available from: Radiopaedia.org.
2. Votruba M, Hungerford J. Primary monophasic synovial sarcoma of the conjunctiva. *Br J Ophthalmol* [Internet]. 2002 [cited 2014 Jul 29];86(12):1453–4. Available from: British Journal of Ophthalmology bjo.bmj.com.
3. Terasaki H, Niki T, Hasegawa T. Primary synovial sarcoma of the lung: a case report confirmed by molecular detection of SYT-SSX fusion gene transcripts. *Japanese J Ophthal* [Internet]. 2001 [cited 2014 Jul 29];**31**(5):212–6. Available from: http://jjco.oxfordjournals.org/content/31/5/212.short.
4. Murphey M, Gibson M, Jennings BT, Crespo-Rodriguez AM, Fanburg-Smith J, Gajewski DA. Imaging of synovial sarcoma with radiologic-pathologic correlation. *Radiographics* [Internet]. 2006 [cited 2014 Jul 29];**26**:1543–66. Available from: www.rsna.org/rsnarights.
5. Shukla P, Pathy S, Sen S, Purohit A. Primary orbital calcified synovial sarcoma: a case report. *Orbit* [Internet]. 2003 [cited 2014 Jul 29];22(4):299–303. Available from: http://informahealthcare.com/doi/abs/10.1076/orbi.22.4.299.17246.
6. Ratnatunga N, Goodlad J, Sankarakumaran N, Seimon R, Nagendran S, Fletcher C. Primary biphasic synovial sarcoma of the orbit. *J Clin Pathol*. 1992;**45**(3):265–267.
7. Oh H, Yang J, Kim J. A Case of primary orbital synovial sarcoma. *J Korean Ophthalmol Soc* [Internet]. 2012 [cited 2014 Jul 29];**53**(5):716–20. Available from: http://synapse.koreamed.org/search.php? where =aview&id=10.3341/jkos.2012.53.5.716&code=0035JKOS&vmode=FULL.
8. Liu K, Duan X, Yang L, Yu Y, Liu B. Primary synovial sarcoma in the orbit. *J AAPOS* [Internet]. 2012 [cited 2014 Jul 29]; **16**(6):582–4. Available from: http://www.sciencedirect.com/science/article/pii/S109185311200328X.
9. Hartstein M, Silver F, Ludwig O, O'Connor D. Primary synovial sarcoma. *Ophthalmology* [Internet]. 2006 [cited 2014 Jul 29];**113**(11):2093–6. Available from: http://www.sciencedirect.com/science/article/pii/S0161642006008542.