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A SPINAL LESION IN A YOUNG ZAMBIAN FEMALE PATIENT: A CASE REPORT

***, [1]Duncan D. Mugala and [2]Yasser Elzanaty**

[1]Senior, Senior Consultant Surgeon Ndola Teaching Hospital, India
[2]Part time Lecturer Copper Belt University-School of Medicine Ndola, India

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ABSTRACT

We have come to know that patients who have this type of illness may come in with Neuroblastomas, Synovial sarcoma tumours Or Chondrosarcomas. The Neuroblastomas. These are CNS tumours and they are termed as neuroectodermal tumours. They are usually phenotypically poorly differentiated, or show divergent differentiation along neuronal, astrocytic and ependymal lines. It is a heterogeneous group of embryonal tumours that occur predominantly in children and adolescents and show aggressive clinical behavior (David N. Louis *et al.,* 2007). More about Neuroblastomas is that although it is a childhood cancer, it can also be inherited in some cases, but the genetic aetiology is largely unknown in (Mossé *et al.,* 2008; Diskin *et al.,* 2009). The Synovial sarcomas are is a rare soft tissue tumor of children and adults that is unrelated to synovium and can occur in almost any part of the body. The familiar biphasic synovial sarcoma has discernible glandular or solid epithelial structures, and monophasic forms have characteristic ovoid or spindle cells with only immunohistochemical or ultrastructural evidence of epithelial differentiation (Mackenzie, 1966). By and large Synovial sarcoma is a high-grade tumor that is associated with poor prognosis. Chondrosarcomas are uncommon, particularly of the bones of the hands and feet but Chondrosarcoma of the pelvis is common, comprising approximately 25% of all chondrosarcomas in most large series. In contrast, chondrosarcoma of the sacrum and mobile spine is rare, constituting less than 5% of all cases (Peter Bergh, 2001). Which one a patient presents with one of these tumours is not an easy diagnosis.

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INTRODUCTION

When we saw this patient our minds were considering several causes of this large rapidly developing tumour causing this problem. These tumors in our minds were by and large three types; we thought of Neuroblastomas (NB), Synovial Sarcoma (SS) or Chondrosarcoma (CS). The First one was likely to be a CNS tumour termed as a neuroectodermal tumour and this type was the Neuroblastoma. The Tumours with only neuronal differentiation are termed CNS Neuroblastomas. This term refers to a heterogeneous group of embryonal tumours that occur predominantly in children and adolescents and show aggressive clinical behaviour. They are usually phenotypically poorly differentiated, or show divergent differentiation along neuronal, astrocytic and ependymal lines.

***Corresponding author:* Duncan D. Mugala**
Senior, Senior Consultant Surgeon Ndola Teaching Hospital, India.

It is a heterogeneous group of embryonal tumours that occur predominantly in children and adolescents and show aggressive clinical behavior (David Louis *et al.,* 2007). A subset of these tumours will undergo spontaneous regression while others show relentless progression (Maris *et al.,* 2007). It is a type of cancer that starts in certain very early forms of nerve cells found in an embryo or fetus. The differences in outcome for patients with neuroblastoma are well known. The patients who have low-risk and intermediate-risk neuroblastoma have a good prognosis and outcome. However, patients with high-risk disease continue to have very poor outcomes despite intensive therapy. The symptoms of neuroblastoma are many. The pain may cause the child to limp, refuse to walk, or become unable to walk. Other symptoms may include the following (Recorded in Neuroblastoma - Childhood: Symptoms and Signs which was Approved by the Cancer.Net Editorial Board, 04/2016)

- A lump or mass in the abdomen, chest, neck, or pelvis, often found by a parent when bathing the child
- Skin lesions or nodules under the skin with blue or purple patches
- Eyes that bulge out and dark circles under the eyes, if the cancer has spread behind the eyes
- Changes in the eyes, such as black eyes, a droopy eyelid, a pupil that is constricted, vision problems, or changes in the color of the iris
- Pain in the chest, difficulty breathing, or a persistent cough
- Pain in the arms, legs, or other bones
- Pain in the back, or weakness, numbness, or paralysis of the legs if the tumor has spread to the spinal cord
- Fever and anemia, which is a low level of red blood cells
- Constant diarrhea or high blood pressure caused by hormones released by the tumor
- Rotating movements of the eyes and sudden muscle jerks, likely from immune system problems caused by the disease

Unfortunately, approximately 70-80% of patients older than 18 months present with metastatic disease, usually in the lymph nodes, liver, bone, and bone marrow (Norman *et al.,* 2016). Around half of all cases are currently classified as high-risk for disease relapse, with overall survival rates less than 40% despite intensive multimodal therapy (Maris *et al.,* 2007). Histologically there are subtypes of neuroblastoma, these are, ganglioneuroblastoma which is an increased schwannian stroma. ganglioneuroma: which is a mature ganglion cell with schwannian stroma. Sometimes the occurrence of neuroblastoma is the appearance of Olfactory neuroblastoma. These are mainly in the nasal cavity; it is the tumor localized in the nasal cavity and paranasal sinuses; and occasionally the tumor extends beyond the nasal cavity and paranasal sinuses. The Olfactory neuroblastoma is radioresponsive in treatment but often it is known to be of a limited extent. By and large the Olfactory neuroblastoma tumor is radiocurable but varies in aggressiveness (Sidney Kadish *et al.,* 1976).

Some other things about neuroblastoma is the association between neuroblastoma and opsoclonus-myoclonus syndrome (OMS) which was described as early as 1927. OMS occurs in 2-3% of patients with neuroblastoma, but neuroblastoma is found in as many as 50% of children who present with OMS. It is believed that OMS is a result of an autoimmune process, not metastases (Rothenberg *et al.,* 2006). More about Neuroblastoma is that although it is a childhood cancer, it can also be inherited in some cases, but the genetic aetiology is largely unknown. Mosse' YP et al identified a significant linkage signal at chromosome bands 2p23-24 using a whole-genome scan in neuroblastoma pedigrees. They used the germline mutations in the anaplastic lymphoma kinase (ALK) gene to explain the hereditary neuroblastomas (Mossé *et al.,* 2008; Diskin *et al.,* 2009). Neuroblastoma (NB) is a tumor derived from the neural crest that usually originates in the adrenal medulla, but it may arise anywhere within the sympathetic nervous system. NB is a common malignancy in children but rarely occurs in adults; less than 10% of all cases are diagnosed after the age of 10 years (Vénat-Bouvet *et al.,* 2010). The second tumour we had in mind was the Synovial sarcoma (SS). These are Synovioblastic origin of the neoplasm these are the results of histochemical staining procedures (Joel *et al.,* 1975). SS tends to affect young people, however, adequate local control may affect survival (Andrew). Synovial sarcoma (SS) is a rare soft tissue tumor of children and adults that is unrelated to synovium and can occur in almost any part of the body. The familiar biphasic synovial sarcoma has discernible glandular or solid epithelial structures, D. H. Mackenzie states that the essential biphasic histological pattern of these tumors are divided into 3 histological groups. Group A includes tumors in which the spindle cell elements and the pseudoepithelial ones are clearly seen throughout the tumor. In Group B the spindle cell (fibrosarcomatous) elements predominate. Group C tumors show mainly a pseudoepithelial structure (Mackenzie, 1966). In this tumour ,the first description of a primary, nonrandom change in the sex chromosome of a human solid tumor is the X;18 rearrangement. It appears to characterize the synovial sarcoma. C Turc-Care et al carried out a translocation that involved chromosome X (band p11.2) and chromosome 18 (band q11.2). This was observed in short-term in vitro cultures of cells from five synovial sarcomas and one malignant fibrous histiocytoma. In four of these tumors, the translocation t(X;18) (p11.2;q11.2) was reciprocal (Turc-Carel). By and large Synovial sarcoma is a high-grade tumor that is associated with poor prognosis. Jonathan J. Lewis, et al, go on to mention that; previous studies analyzing prognostic factors are limited because of inclusion of heterogeneous cohorts of patients with nonextremity and recurrent tumors. In their objective study they tried to determine independent prognostic factors of primary synovial sarcoma localized to the extremity. They concluded that the natural history of primary synovial sarcoma of the extremity is related to tumor size and invasion of bone and neurovascular structures (Jonathan *et al.*).

The third likely tumour in our mind was that of Chondrosarcoma (CS). We have come to know that Chondrosarcomas of the bones of the hands and feet are uncommon. David C. Dahlin and Antonio H. Salvador wrote up their review of 30 lesions revealing that, histologically, Chondrosarcomas may be difficult to distinguish from benign cartilaginous tumors of soft tissue or of bony origin. They go on to say that Roentgenographic features and the operative findings may afford critical evidence for the nature of the disease. Pain is commonly present, in contrast with its rarity in chondromas (David *et al.,* 1974). We also know that Chondrosarcoma of the pelvis is common, comprising approximately 25% of all chondrosarcomas in most large series. In contrast, chondrosarcoma of the sacrum and mobile spine is rare, constituting less than 5% of all cases (Peter Bergh *et al.,* 2001). Chondrosarcoma of the synovium, either primary or secondary to synovial chondromatosis, is rare. We also discovered that chondrosarcomas originating in the facial skeleton have been reported. However, the information regarding chondrosarcomas in the facial region is very rare. The other issues about Chondrosarcomas is that most chondrosarcomas of the pelvis, sacrum, and spine are considerably larger than their appendicular counterparts, averaging 11 cm in size at diagnosis. Peter Bergh et al go on to state that surgical options are limited or dictated by the tumor's proximity to vital structures as well as the risk of jeopardizing axial stability (Peter Bergh *et al.,* 2001).

CASE REPORT

In June 2017, we met GC a female 16 years old who was healthy but in severe pain on her left side of abdomen

extending to the upper left thigh. The tumour had grown rapidly within one and half months since the illness developed (45 days). She came with flexion deformity of left hip joint. Physical examination revealed an abdominal mass there was a palpable mass on left iliac fossa, with close contact with left inguinal ligament (inferior) and ASIS (lateral), extending medially to umbilicus and to left costal margin (superior), It was firm and fixed, the rest of the abdomen was normal. She had severe pain during an attempt to passively move the left hip joint extension, the left femoral artery was palpable,

There was no fever, The patient opened bowels normally, there were no urinary symptoms, She was menstruating with no gynecological complaint.

The Abdominal Ultra Sound scan:

To determine the mass was done, but the original of the mass was not clear, the CT Abdominal-pelvic was ordered but not done. Family could not afford it. The decision for Exploratory laparotomy was done. A laparotomy was carried out and clinically, the diagnosis of neuroblastoma was made. We suspected NB and knew that currently, there were no standard treatment guidelines for such young patients. In such patients with metastatic disease with a late stage of disease, multimodal therapy should include surgical resection, radiotherapy, and outpatient chemotherapy**.** The decision for the exploratory laparotomy was done.

Operative Finding were:

- Lower midline incision was done, opening the peritoneum. The mass found was in retroperitoneal on left gutter, near the mobilization of sigmoid colon done with access to left retroperitoneal space through white line of Toldt, the left ilio-poas muscle was pushed interiorly with the mass, retracting the muscle medially done and the left ureter seen over the muscle.
- The mass was very fixed to underlying Qaudratus Lamborum muscle and to lumbar vertebrae medially with close relationship with aortic bifurcation and abdominal aorta.
- Blunt dissection of the mass was done with removal piece by piece. and haemostasis with abdominal towels inserted. Then complete excision was achieved. And packing the raw area of mass origin with surgicel haemostatic.

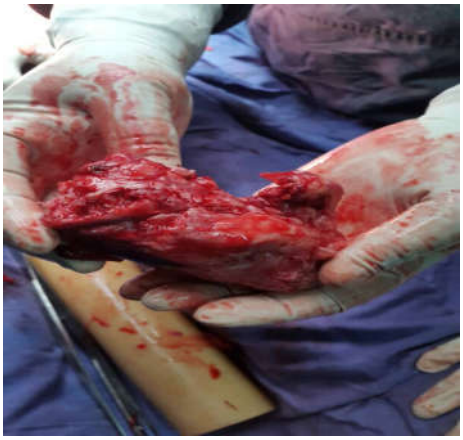

Figure 1. Neuroblastoma or fibroma

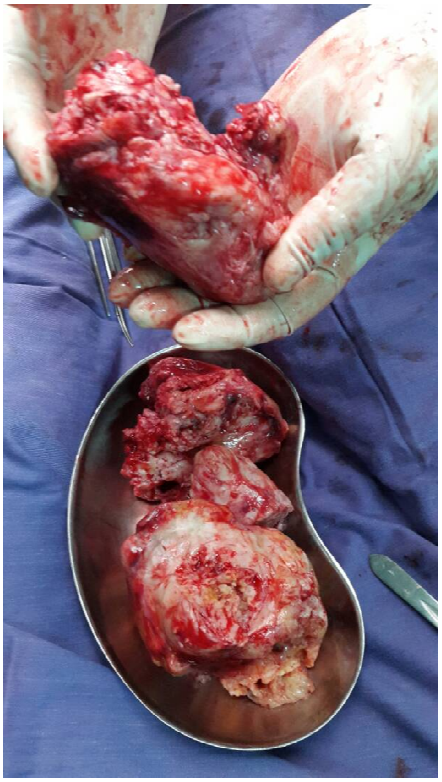

Figure 2. Neuroblastoma or fibroma

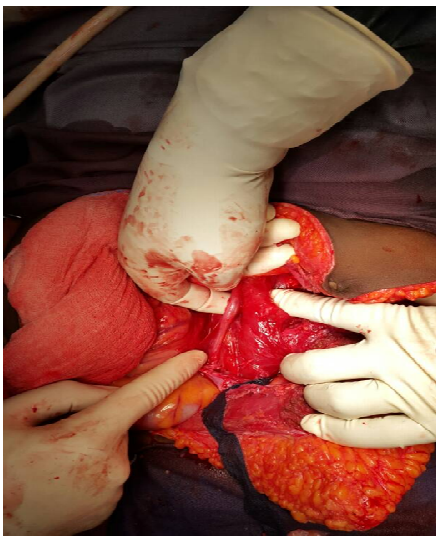

Figure 3. Ureter

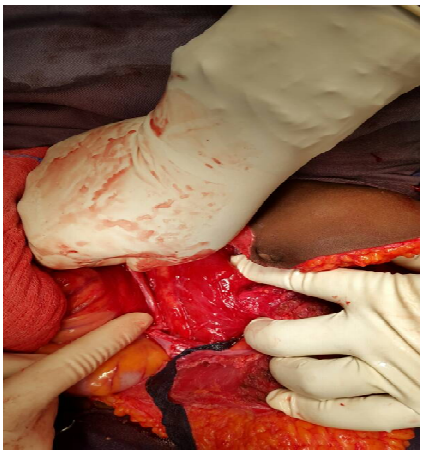

Figure 4. Ureter Isolated

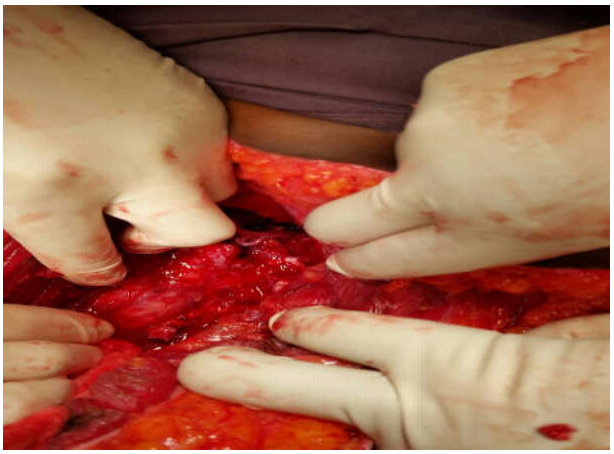

Figure 5. Where the Tumour was excised

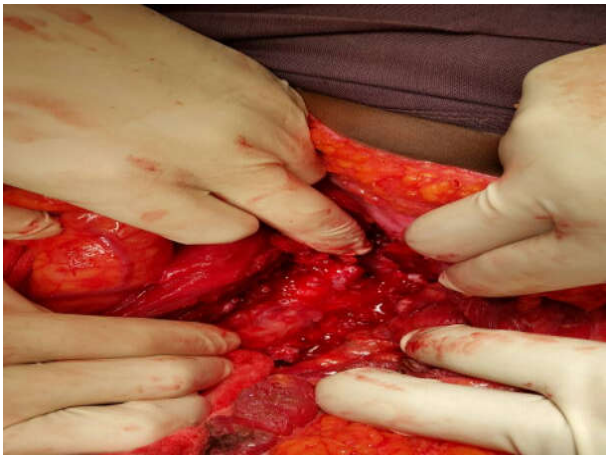

Figure 6. Where the Tumour was excised

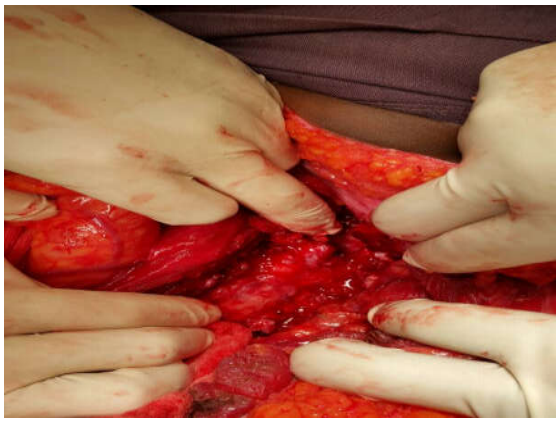

Figure 7. Where the Tumour was excised a about T12 to L2

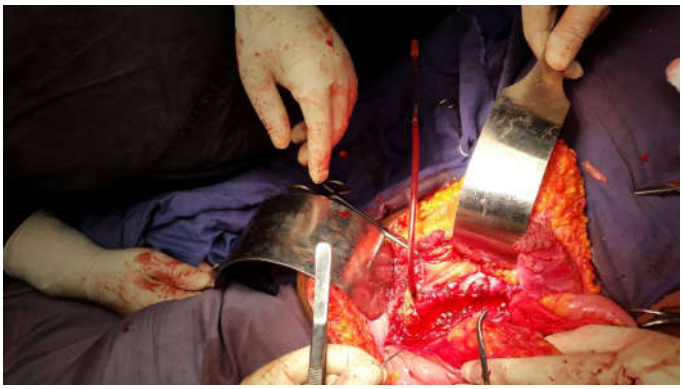

Figure 8. Wound sutured. A drain tube in line

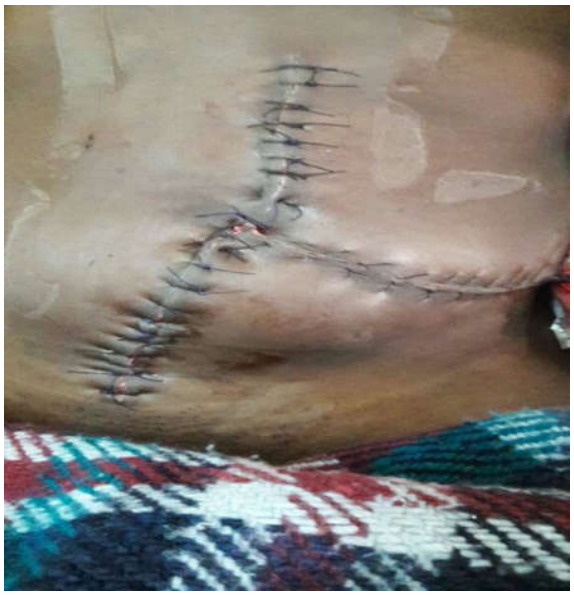

Figure 9. Sutured abdomen

DISCUSSION

The differences in outcome for patients with neuroblastoma are striking. Patients with low-risk and intermediate-risk neuroblastoma have excellent prognosis and outcome. However, those with high-risk disease continue to have very poor outcomes despite intensive therapy. Unfortunately, approximately 70-80% of patients older than 18 months present with metastatic disease, usually in the lymph nodes, liver, bone, and bone marrow. Less than half of these patients are cured, even with the use of high-dose therapy followed by autologous bone marrow or stem cell rescue (Norman, 2016). Our patient developed malignancy at the age of 16 years. It was a bone type of the tumour. We suspected NB in this young lady in that less than 10% of all cases of NB are diagnosed after the age of 10 years (Vénat-Bouvet, 2010). In our mind she was in a high risk patient in that The symptoms of neuroblastoma are many. The pain may cause the child to limp, refuse to walk, or become unable to walk. Other symptoms may include the following (Recorded in Neuroblastoma - Childhood: Symptoms and Signs which was Approved by the Cancer.Net Editorial Board, 04/2016). Our patient was in a state of being unable to walk and was in severe pain in the left lower limb (Norman, 2016 and Maris, 2007). More about Neuroblastoma is that although it is a childhood cancer, it can also be inherited in some cases, but the genetic aetiology is largely unknownIn. Mosse' YP et al identified a significant linkage signal at chromosome bands 2p23-24 using a whole-genome scan in neuroblastoma pedigrees. They used the germline mutations in the anaplastic lymphoma kinase (ALK) gene to explain the hereditary neuroblastomas (Mossé, 2008 and Diskin, 2009). In adults neuroblastoma tumours are significantly worse than in children. This may be due to tumor biology, more virulent clinical course, or possibly due to the fact that adults are less sensitive or have poor tolerance to pediatric chemotherapy regimens. Clinical data on survival outcomes of adult patients (defined as 20 years of age and older) with neuroblastoma are scarce due to the rarity of the disease. Our patient was 16 years old and was by and large close to the 20 year old stage. Small single institution reports have described worse outcomes for adults than pediatric patients (Esiashvili, 2007). We came to know that her grandmother had a similar tumour on the left

side of abdomen and the tumour was very aggressive and she died. She was almost 90 years old. Generally it is known that Neuroblastoma (NB) is a rare malignancy in adults, it is a documented occurrence and can have grave implications for patients. However adult and pediatric patients with neuroblastoma achieve similar survival outcomes (Henry, 2014). The second tumour we had in mind was the Synovial sarcoma (SS). These are Synovioblastic origin of the neoplasm these are the results of histochemical staining procedures (Joel, 1975). SS tends to affect young people,and their survival is not good. The third likely tumour in our mind was that of Chondrosarcoma (CS). We have come to know that Chondrosarcomas of the bones of the hands and feet are uncommon. We also know that Chondrosarcoma of the pelvis is common, comprising approximately 25% of all chondrosarcomas in most large series. In contrast, chondrosarcoma of the sacrum and mobile spine is rare, constituting less than 5% of all cases (Peter Bergh, 2001). Out patient's tumour was largely in the sacrum.

Conclusion

A sample was taken and the report came to us and it revealed the diagnosis was the Synovial sarcoma (SS). Despite our effort for this young lady to go for chemotherapy and or radiotherapy she died after a period of two months from the time she was operated on.

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