Osteosarcoma: A Literature Review and Case Study

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Abstract

Osteosarcoma is a disease that mainly affects the long bones of the body and arises during times of rapid growth. Conversely, osteosarcoma can arise in short bones and can occur during later stages of growth. The sign of osteosarcoma on a radiograph gives a sunburst appearance. It can also be seen with computed tomography scans as well as with magnetic resonance imaging. There are different types of treatments, but the majority of options include chemotherapy, surgery, amputation, replacements or some type of prosthetic.
Introduction

Osteosarcoma can affect any person at any age. It occurs mainly in long bones but has been known to occur also in short bones. Mainly, children are affected because of the rapid growth, but that does not mean they are the only ones affected; adults can also be affected. Osteosarcoma can be treated with chemotherapy, surgery, prosthetics, amputations, or rotationplasty. Follow-up care is necessary in case of relapse and to monitor patient’s progress.

Literature Review

Bone Sarcoma

Osteosarcoma is the most common primary sarcoma of bone. Osteosarcoma occurs mostly during puberty or periods of rapid bone growth, but it can also occur in younger children as well as in older adults. According to Findik et al. (2012), “Osteosarcoma is classified as a component of Ewing’s sarcoma…generally [has] long bone involvement such as the femur, tibia and humerus…very rarely [it] arises from the short bones such as [the] ileum, clavicle, scapula and sacrum” (p.173). With osteosarcoma being rare in short bones, such as the clavicle and scapula, it has also been known to occur in the head and neck. However, instances in the head and neck “account for less than 10 percent of all osteosarcomas in general” (Yamamoto et al. 2011, p. 201). Gebhardt, Dempsey, and Neff (2008) state that osteosarcoma “can arise in the flat bones of the pelvis, skull, scapula, and ribs and in the spine. Overall, the majority of the lesions develop in the extremities and pelvis” (p. 1948).

Since osteosarcoma can arise in many different places in the body, there are different ways to detect and treat it. Learning to recognize the signs and symptoms of osteosarcoma is one of the first steps in diagnosing the disease. According to Babazade, Mortazavi, and Jalalian (2011) and Guillon et al. (2011), the most common symptom is pain. If a patient does not present
with pain or complain of any pain, osteosarcoma is usually diagnosed because of a traumatic event; it is discovered by chance. Noticing how osteosarcoma appears on a radiograph is the next step to recognizing the problem. Gebhardt et al. (2008) states, “The plain radiograph is the best diagnostic tool. Osteosarcomas may either completely destroy the bone (radiolucent lesion) or replace the bone with a blastic response (radiodense), but they most often do both” (p. 1951).

Once discovered, options for treatment depend on how badly the disease has taken over the bone.

**Chemotherapy**

Chemotherapy is used to treat osteosarcoma. It can be used as neoadjuvant chemotherapy and post-operative chemotherapy. Neoadjuvant chemotherapy can be necessary when a physician is looking to perform surgery on the affected area. The chemotherapy helps make the tumor smaller, so surgery will not have to be as drastic as first imagined. It also gives the patient and the physician time to understand what needs to be done and to communicate in a way that will benefit the patient and their lifestyle.

According to Hayden and Hoang (2006), osteosarcoma can be divided into different subtypes. There is conventional osteoblastic osteosarcoma and it makes up 70% of all osteosarcomas. There is also chondroblastic and fibroblastic which make up 10% each. The less common types are anaplastic, telangiectatic, giant-cell rich, and small cell. The fibroblastic subtypes have the best response to chemotherapy and the chondroblastic subtypes have the worst response. Hayden and Hoang (2006) state that, “several mechanisms are responsible for resistance to chemotherapy. [For example,] P-glycoprotein…is an ATP-dependent efflux pump that may confer resistance to doxorubicin and etoposide” (p.4). Doxorubicin and etoposide, along with methotrexate, ifosfamide and cisplatin, are medications that are used in neoadjuvant
and post-operative chemotherapy (Hayden & Hoang, 2006; Gebhardt et al., 2008). With the help of chemotherapy, osteosarcoma responses have improved.

**Surgical Options**

According to Gebhardt et al. (2008), adjuvant chemotherapy was first introduced in the 1970s and is now established as an essential part of treatment of osteosarcoma. It used to follow amputation or resection of a tumor. They state that, “The use of neoadjuvant chemotherapy [allows] the assessment of histologic response to the chemotherapy, which has prognostic implications, allows time for planning of limb salvage operations, and makes those resections easier and probably safer” (p.1953-54). It can also help give the surgeon time to make a prosthetic if one is needed, and it gives him time to work with the patient and prepare him/her for surgery and post-operative procedures such as post-operative chemotherapy and physical therapy, if needed. The surgical treatment of osteosarcoma is complete resection of the tumor. Surgery is a helpful option for patients, but it does not help patients deal with the fact that they could lose a limb, look deformed, or possibly retain their limb but not have full use of it.

According to Grimer (2005), rotationplasty is the “removal of a diseased portion of bone, turning the shortened leg bone through 180°, and reattachment of the limb” (p.89). Grimer (2005), continues to explain that in “othopaedic oncology, the technique has been modified to allow resection of a tumour around the knee by attachment of the healthy lower leg and foot to the upper thigh after rotation through 180° and preservation of the sciatic nerve” (p. 89). (See Figure 1) Doing so allows the patient’s foot to act as the knee joint once it is fitted with a prosthetic. Grimer (2005), also says, “the advantages of [the] reconstruction are improved function, a fairly low risk of complications, and absence of phantom pain.” (See Figure 2)
Grimer (2005) says that “for many patients and families, amputation is seen as a failure of treatment” (p.90), but Gebhardt et al. (2008), see it as an option. Gebhardt et al. (2008) say that “we have become so accustomed to salvaging limbs that amputations are probably not being used enough…[and] with modern prosthetics for the lower extremity, function can be quite good….Amputation should not be viewed as a failure or sign of defeat” (p.1965). Amputation can be difficult for patients because of the fact that they lose a limb. Patients also are faced with the fact that they look different and people will stare or possibly make fun of them. This need not be the case because of the technology that is available today. Prosthetics have improved and patients can live a more normal life than they would have been able to when prosthetics were first being used.

**Examining Different Studies**

Short bones are still affected by osteosarcoma even though it mainly occurs in long bones. Findik et al. (2012) discuss a case pertaining to osteosarcoma of the rib. The patient was a 28-year-old woman who had left-sided chest pain for 3 months. She had a chest x-ray that showed opacity on her left side near the sixth and seventh ribs. She had a computed tomography (CT) scan of her thorax as well as a positron emission tomography (PET) fluorine 18 fluorodeoxyglucose scan and the scan depicted that the lesion was a low-grade malign neoplasm. She underwent a thoracotomy and, after being discharged, she was referred for chemotherapy. Upon her 17th-month follow-up she had remained healthy and was disease free.

There are some similarities when comparing small cell osteosarcoma of the rib to osteosarcoma of the mandible. Babazade et al. (2011) point out that osteosarcoma of the jaw is rare and makes up 5% to 13% of all cases of osteosarcoma. This particular case was a 27-year-old male with painful swelling in the right mandibular body. After a CT scan, an osteolytic lesion
was discovered over the right mandibular body. It involved both the inner and outer cortices, and it also made a sunburst appearance. (See Figure 3) The patient had a resection of the mass, and his jaw was reconstructed with a plate for the right side of his mandible. After surgery, he underwent post-operative chemotherapy. Thirteen months later, the patient noticed another painful mass on the left mandibular body, and after a CT scan there was another sunburst appearance. With the next surgery, the previous plate that had been put in place was removed and replaced by a longer plate that included the condylar process. (See Figure 4) Again, chemotherapy was repeated, and a month after his second surgery the patient was doing well.

Comparing these two cases shows a few similarities. Both cases are rare occurrences of osteosarcoma because they occurred in short bones: the rib and the mandible. Both patients underwent chemotherapy after their procedures, but the difference was that the male experienced a second occurrence on the opposite side of his jaw while the female remained healthy even into the 17th month following her thoracotomy. When a recurrence occurs, it is usually within five years after treatment. In the case of the male, it was just shortly over a year. After a recurrence the chance for another relapse is even greater. Both cases are rare, but they show that osteosarcoma can affect short bones and can affect both males and females. It also shows that recurrences happen but that they can be treated.

Osteosarcoma arising from short bones is rare, but osteosarcoma arising from osteochondroma is even more rare. Engel et al. (2012) discuss a case in which a 13-year-old male underwent two different resections for osteochondroma, and upon further evaluation for a new complaint, the osteochondroma had evolved into osteosarcoma. The patient had complained of pain and swelling on the left proximal tibia. The patient underwent a partial resection, and four years later the patient then complained of joint limitation and pain. In the previous resection,
spiculous bone formations caused a prominence in the anteromedial aspect that limited knee movement. The patient underwent another partial resection. Eight years later, the patient was complaining of pain and noticed there was a bulge on the postero-medial aspect of the tibia. The histology of the bone that had been resected showed that it was osteosarcoma.

Looking at the first and second specimens from the resected bone, the findings were similar, but when compared with the third specimen it appeared different. The first two specimens “had a cap of firm, gray, translucent cartilage”, and the third specimen was irregularly shaped and had “bony excrescence that projected from the surface of the tibial bone, and was partially covered by a bluish cartilaginous cap” (Engel et al., 2012, p.450). It was concluded that the tumor was a low-grade osteosarcoma. When looking at this case, it shows that osteochondroma transforming into osteosarcoma is rare, and it is estimated to have an occurrence of less than 1-3%. It also shows that when a disease is diagnosed, it does not always remain the same.

**Follow-Up Care**

Kaste (2011), states, “approximately 30% of patients with osteosarcoma will develop disease relapse, most often involving the lungs” (p.759). Hayden and Hoang (2006) support that data and add that pulmonary metastasis is the most common form of spreading, which occurs in 80% of patients, and bone involvement is the second most common at 15%. Patients with any type of bone sarcoma are monitored for at least 5 years after diagnosis, and the Children’s Oncology Group Bone Tumor Committee actually suggests that patients have 10 years of follow-up to monitor primary and metastatic diseases (Meyer et al. 2008).

**Case Report**
During an open reduction internal fixation (ORIF) procedure on a 48-year-old male, osteosarcoma was diagnosed. The patient underwent chemotherapy, after which a total humerus replacement was performed. The patient’s main symptom was upper arm pain, and there was nothing in his family history or medical records to give reason to have suspected osteosarcoma. The patient was referred to a physician for a pathological fracture and metastatic bone tumor. At that time, the patient had swelling and heat present in the proximal forearm. He also experienced pain and tenderness during motion.

When radiographs were obtained, there was a pathological fracture in the proximal humerus, “showing hypertranslucency of bone, mainly involving osteolytic changes” (Yoshida & Tokuhashi, 2012. p.1) (See Figure 5). After images were acquired from CT, magnetic resonance imaging (MRI) and bone scintigraphy, findings indicated a pathological fracture with a metastatic bone tumor. An ORIF was performed and 3 ender’s pins were inserted at the elbow joint. Following treatment severe swelling occurred. After pathological examination, the diagnosis was an osteoblastic-type sarcoma. Yoshida and Tokuhashi (2012) discuss what the patient went through after he achieved a complete response to the preoperative chemotherapy.

[The patient underwent] massive extra-capsular resection…followed by total humerus replacement…The humerus was entirely resected including parts of the deltoid and triceps brachii muscles and cuff, whereas the long head of the biceps brachii muscle, musculocutaneous nerve, and other neurovascular bundles were conserved. In reconstruction of the cuff, an artificial ligament was attached around an artificial bone head…the artificial joint was entirely covered with a latissimus dorsi muscle flap (p. 2). (See Figures 6 and 7)
After surgery the patient underwent postoperative chemotherapy, and eight years after surgery the patients condition was still favorable and was able to resume his normal activities. The only problem he had was difficulty contracting his elbow even after 8 years had passed since the surgery.

Discussion

Osteosarcoma is shown to affect anyone at any age. There are different types of osteosarcoma with some being more prevalent than others. Osteosarcoma can also become apparent after having received treatment for a different type of sarcoma. Even though osteosarcoma mainly affects long bones, short-bone involvement is also possible. Though the latter is rare, it needs to have attention brought to it so it can be caught early and have treatment administered. The different treatments need to be discussed with the patient. Neoadjuvant chemotherapy, surgery, and post-operative chemotherapy are the main treatments. With surgery, the actual process needs to be explained to the patient so they understand what needs to be done. Whether it is a limb resection, rotationplasty, a replacement or an amputation, the patient needs to understand why a given treatment is selected and what options they have after the surgery. Patients also need to understand the follow-up care because of the recurrence of osteosarcoma. The patient needs to understand that once they are treated, the sarcoma can still return in another part of the body. Regular check-ups need to be followed to monitor any changes. Osteosarcoma is the most common sarcoma of the bone, but it does not mean that once a patient is diagnosed with it there is not any help. It does not mean that once they are diagnosed there is no chance for treatment. They just need to know what their options are, what they can do to help the process, and the care they need after treatment.
Figures

**Figure 1.** Reattachment of the lower limb during rotationplasty surgery.


**Figure 2.** Rotationplasty after surgery and with the prosthesis.

**Figure 3.** CT scan showing a sunburst appearance of the right mandibular body.

![CT scan of the right mandibular body](image)


**Figure 4.** Final panoramic image after the second surgery.

![Panoramic image of the mandible](image)

**Figure 5.** MRI image of the right proximal humerus

![MRI image of the right proximal humerus](image)


**Figure 6.** Image taken during ORIF right humerus surgery.

**Figure 7.** Postoperative radiograph after ORIF of right humerus.
References


