INTRODUCTION

Tumors of the hand are uncommon, and malignant tumors of the hand are particularly rare. Most of the tumor types encountered in bone and soft tissue in other body regions also occur in the hand, but the frequency and distributions are different.[1-3] Tumors involving the hand are classified as benign, malignant and metastatic. Benign tumors are the most common tumors in the hand and are classified as latent, active, and aggressive.[4] Enchondromas are the most common and destructive primary bone tumors of the hand skeleton. Malignant osseous neoplasms of the hand are so rare that even single case warrants publication. The hand may be the site of distant breast, kidney or lung metastases, most of which occur in the distal phalanges. Because the hand has limited free space and exquisite sensitivity, however, even small, histologically benign masses can cause pain, impairment of function, or obvious swelling.[5]

Tumor like conditions in the hand are frequent such as ganglion cyst, epidermal inclusion cysts, sebaceous cyst. The ganglion cyst is the most common soft tissue lesion of the hand and wrist.[6]

Independent of tumor type, most of the patients with hand tumors present with pain and/or mass. Clinical findings can be nonspecific in metastatic disease, and can include the general signs of inflammation (pain, swelling and erythema).[1]

Pathologic fractures can occur with benign lesions such as enchondroma, and with malignancies such as chondrosarcoma. For a few tumor types, clinical presentation can be helpful in diagnosis. Pain due to osteoid osteoma is characteristically relieved by aspirin.[7] Lesions located on tendons suggest tumors of tendinous origin, such as giant cell tumor of the tendon sheath. A purple lesion under the nail accompanied by pain with no history of trauma is suggestive of glomus tumor.[8]

Imaging generally provides a preliminary basis for deciding whether a lesion is benign or malignant. With bone tumors in the hand, as in other body regions, sclerotic margins suggest a benign lesion, while a lack of margins or a breakdown of cortex suggests malignancy. Bony changes are more apparent on CT, and in particular any intramedullary involvement can be evaluated for preoperative planning of the extent of resection that will be needed. Soft tissue lesions can be shown in greater detail with MRI. Needle or surgical biopsy may be necessary in providing a precise diagnosis for treatment planning.[1,9]

For patients with benign lesions in bone, the main indication for surgery is pain. Benign lesions in soft tissue may require surgical treatment if they are locally aggressive or if they interfere with hand function. For aggressive benign bone tumors such as giant cell tumor, the use of adjuvants with curettage has been recommended.[10] However, for giant cell tumors in the finger, amputation may be necessary.[11]

For lesions that are suspected of being malignant, histopathologic diagnosis is needed before surgery. Obtaining a wide surgical margin may require amputation. For example, with tumors of the finger, when extracompartmental extension or pathologic fractures are present, removal of the finger or ray resection is indicated.(12)

In metastatic disease, definitive treatment depends on finding the primary malignancy and treating the patient accordingly.(13)

Due to the hand's complex structures being located in a small space, the risk of inadequate surgery that preserves structure and function must be balanced against the risk of unnecessarily extensive surgery.(14)

AIM OF THE STUDY

This study aims to discuss various types of osseous and soft tissue tumors and tumor like conditions of the hand and its management.

CLINICAL DIAGNOSIS

History

A thorough history and physical examination remain the cornerstones of initial patient assessment. The patient should be asked the duration of symptoms, swelling, or mass. A history of pain or recent growth may give information regarding the aggressive potential of the lesion. It is important to remember that soft tissue sarcomas are often painless. These lesions may be present for long periods with little or no growth. History of exposure to radiation, herbicides, or other carcinogens may help in generating a differential diagnosis. The family history may provide insight into hereditary diseases or genetic syndromes that predispose to sarcoma. [15,16]

Physical Examination

Physical examination includes the assessment of the lesion in question, including swelling, color, size, warmth, consistency, tenderness, mobility, and proximity to fascia, tendons, nerves, and vessels. Transillumination can demonstrate fluid or myxomatous tissue, as seen with a ganglion or soft tissue myxoma. The location of the lesion is important in formulating a differential diagnosis. If a diagnosis of ganglion cyst is considered, the lesion should be in a typical or well-described location. An unusual location should raise the suspicion of an alternate diagnosis. The nerve and blood supply

to the hand should be carefully assessed. Regional lymph nodes should be initially assessed by palpation and, subsequently, with radiographic studies or biopsy, if indicated.[17]

Clinical presentation

The majority of patients with soft tissue tumors present with a chief complaint of a painless mass. or neurologic symptoms. It is not uncommon for a hand sarcoma to be initially mistaken for a more common disease of the hand.[18,19,20]

Carpal tunnel syndrome has been reported to result from synovial sarcoma, epithelioid sarcoma, giant cell tumor of tendon sheath, and chondrosarcoma.[21,22] It is commonly taught that an important symptom to elicit in the history of present illness in the evaluation of a patient with musculoskeletal pain is night pain, which suggests pain of nonmechanical etiology (i.e., tumor or infection). However, it is well known that patients with carpal tunnel syndrome often have discomfort and pain in the hand at night, so that the history is less helpful in determining if hand symptoms are a result of sarcoma. Sarcoma of the carpal canal does occur, but is a rare cause of carpal tunnel syndrome. Unless an obvious mass is present, one must be aware that occult tumors can present as more common hand syndromes, and when encountered intraoperatively, a biopsy should be properly performed.

The duration and time course of symptoms are also important. Children with a mass present since infancy most likely have hamartomas, hemangiomas, or infantile fibrosarcoma.[23,24] Rapidly growing lesions usually represent sarcoma or inflammation, but can occasionally be a benign lesion like nodular fasciitis, which is a rapidly growing benign lesion commonly misdiagnosed as a soft tissue sarcoma.[25] A history of trauma or penetrating injuries may suggest a foreign body granuloma, epidermoid cyst, or traumatic aneurysm as the etiology of a mass. In older patients, particularly those with a significant history of tobacco use or known history of lung carcinoma, metastatic disease from bronchogenic carcinoma of the lung to the hand is a possibility. Although metastases below the elbow are rare, osseous metastases with a soft tissue mass are usually from lung or renal cell carcinoma.[26] Swelling of the dorsal wrist, volar wrist, and proximal digital crease are common locations for ganglion cysts, by far the most common lesion causing swelling or a mass.[27] Swelling over a distal phalanx is often due to a mucous cyst. The second most common soft tissue tumor in the hand is giant cell tumor of tendon sheath, or pigmented villonodular synovitis, which originates from the synovium of a tendon sheath or joint. Fluctuation of size with palpation and position of the limb is characteristic of hemangioma. If abnormal pigmentation is present, melanoma, nevi, or squamous cell carcinoma (which typically ulcerates) should be suspected. Persistent lesions demonstrating characteristics of a callus (firm, woody hard with

or without ulceration) must be evaluated for epithelioid sarcoma, viral wart, pyogenic granuloma, squamous carcinoma, and sweat gland tumor and other tumors of the skin and appendages. Lesions should be assessed for pulsation and bruit to rule out aneurysm. A patient with pain under the nailbed, cold intolerance, and exquisite tenderness, with or without a mass and purple patch, should be evaluated for a glomus tumor. Epitrochlear and axillary lymph nodes should be palpated in all patients with a potential soft tissue sarcoma because epithelioid sarcoma, embryonal rhabdomyosarcoma, synovial sarcoma, and angiosarcoma are at increased risk for lymph node metastasis.[28]

INVESTIGATIONS

Laboratory Studies

Laboratory studies include determination of levels of serum calcium, phosphorus, blood urea nitrogen, and creatinine to evaluate the possibility of metabolic bone disease. The serum alkaline phosphatase and lactase dehydrogenase levels are elevated in some malignancies such as osteosarcoma and ewing's sarcoma. A serum immunoelectrophoresis assists in determining if multiple myeloma is present. The hematologic profile and the erythrocyte sedimentation rate are abnormal in the presence of many neoplastic and infectious processes. A urine analysis may detect an occult renal cell carcinoma. Antinuclear antibodies and rheumatoid factor may be positive in patients who have upper extremity swelling secondary to rheumatoid disease.[29]

Radiological studies

A variety of diagnostic imaging techniques permit analysis and localization of the size and extent of the lesion and its impingement on normal anatomy. Although imaging does not typically offer a specific diagnosis, it provides important clues in the evaluation, analysis, and work-up of a specific patient.

Plain Radiographs

Plain radiographs afford the best possible resolution and detail of bone and adjacent soft tissues. The anatomic region in question should be fully visualized on all radiographs, and this is achieved by taking an adequate number of views. Plain radiographs are the standard for predicting the presence and location of bony involvement. They may also be the most specific radiologic test for bone tumors.

Computed Tomography

Although conventional radiographs offer significant spatial information, the higher resolution of CT produces images that are useful specifically in localizing small tumors within a bone and in identifying soft tissue extension or calcification within such growths. Because of its ability to discriminate varying densities, a CT scan allows visual separation between the medullary canal, cortex, and surrounding soft tissues, often providing critical information concerning anatomic location (T) for tumor staging. Plain chest radiographs are always obtained before performing a biopsy of a suspected malignancy; chest CT scans are useful for staging histologically defined malignant tumors but are not routinely used for prebiopsy screening studies.

Scintigraphy

Radionuclide "bone scan" imaging can be very helpful in detecting primary and metastatic tumors. However, the phenomenon of increased radiopharmaceutical pooling (increased uptake) in a particular bone or soft tissue area is not at all specific or diagnostic. The technique has two time-variable phases. Within minutes after injection, conditions associated with an increased vascularity show abnormally high uptake (trauma, infection, and neoplasia). Later, the isotopes are actively concentrated; in the skeleton, pooling occurs in woven (new) bone, so that any process that forms immature bone would be associated with increased uptake on films done 2 or 3 hours after injection. Radioisotope scans are probably most helpful to demonstrate lesions that may not have been suspected clinically or in anatomic sites not seen on initial images that focused only on the symptomatic region. These findings may be important in patient management, and they have the potential to alter biopsy or therapeutic plans significantly.

Magnetic Resonance Imaging

MRI is a superior imaging technique for the assessment of the soft tissue extent of disease and is particularly beneficial in the evaluation of soft tissue sarcoma. The extent of intramedullary bone involvement can also be readily determined. This facilitates preoperative planning of bone

transection level during resection and may aid in the identification of skip metastases that otherwise might not be identified. High-resolution CT offers the benefit of more accurate assessment of cortical bone involvement and may be the preferred technique for evaluating lesions involving the cortex of a given bone. CT is particularly beneficial in identifying occult osteoid osteomas.[30,31,32]

Sonography

Sonograms produce images from a differential pattern of transmitted and received echoes. Compared with CT and MRI, sonography produces less detail with respect to precise anatomic relationships and tumor margins. Echo patterns of solid masses are nonspecific, but fluid-filled lesions can be easily differentiated from solid tumors. Sonograms are extremely inexpensive in comparison to the cost of CT and MRI and may be more useful than either for distinguishing reactive and cystic processes

Biopsy

The biopsy is the last stage of diagnostic management. Biopsy surgery should be planned as carefully as the definitive operation. The exact technique used is influenced by history, location, and size of the mass, as well as the experience of the surgeon and the pathologist.

Needle Biopsy

Needle biopsy has an extremely limited role in the diagnosis of lesions in the hand and upper extremity. It can be useful in confirming the histology of a recurrent or metastatic lesion. Needle biopsy produces only a small and often fragmented tissue sample that may be impossible to diagnose or grade accurately. Even if the core biopsy seems clearly diagnosable, the tissue volume may not be representative of the tumor, leading to the possibility of overgrading or under-management.

Open Biopsy

Open surgical biopsy is a complex procedure, but one that plays a critical role in determining treatment outcome for aggressive and malignant tumors. During the biopsy, the patient and surgeon must be prepared for *the unexpected. Institutions or clinicians that are not prepared or able to* complete all diagnostic studies and also provide definitive surgical and medical-adjunctive management are best advised to refer patients before the biopsy is performed.

Hand surgery is most safely and efficiently performed in a bloodless field. The use of a pneumatic tourniquet is acceptable during both open biopsy and surgical treatment of tumors and neoplasms. Limb exsanguination before biopsy is contraindicated before tourniquet inflation because of the risk

of seeding or dislodging tumor cells. An entirely satisfactory field is achievable by elevating the arm for 3 to 4 minutes before inflating the tourniquet. Likewise, the use of intravenous anesthesia (Bier block technique), a method that also requires pretourniquet exsanguination, is not appropriate for tumor biopsy or treatment of aggressive bone or soft tissue neoplasms.

A frozen section is necessary at the time the biopsy is performed to determine whether an adequate specimen has been sampled, so that permanent light microscopic and special microscopic evaluations can be carried out subsequently. It is rare to find a pathologist who can consistently provide accurate histologic analysis of a frozen specimen dependable enough to begin treatment. The definitive treatment plan should be decided on only after the results of all permanent sections, electron microscopic studies, and special tissue techniques are complete and reported. All biopsy specimens should be cultured, and all cultures should be sampled. Although plain chest radiographs are taken before biopsy, chest CT is performed to stage a diagnosed lesion. If the tumor is aggressive or malignant, the biopsy tract itself will be contaminated with tumor cells and must later be excised en bloc with the subsequent resection or amputation specimen. Preferred biopsy incisions are therefore longitudinal and are carefully placed to permit their complete excision without having to extend a dissection margin simply to accommodate a badly placed incision. As a simple rule, the biopsy should be

placed in line with or immediately parallel to the incision that may be required for later attempted limb salvage Transverse and Bruner-type incisions are specifically to be avoided.

Incisional Biopsy

Incisional biopsy is frequently the most appropriate technique for diagnosis of bone and soft tissue masses. It requires excision of an adequate tissue sample that is minimally manipulated. Frozen sections are important to determine specimen sample adequacy. The biopsy incision is located to afford the most direct route to the tumor, thereby ensuring that the fewest tissue planes are disturbed or contaminated.

Correct surgical technique for biopsy is different from all typical surgical dissections. Biopsy involves a direct approach via a longitudinal incision through muscle and other tissues that overlie the mass. Biopsy technique does not include spreading or extensive and vigorous retraction, which has the potential to contaminate a widened area. However, adequate visualization during biopsy, as in any operation, is critical. Because the biopsy tract is cell-contaminated, an approach through a muscle sacrifices only one plane, whereas spreading techniques (i.e., moving muscles aside) contaminate not only the muscle that is retracted but also all surrounding structures. Perfect postsurgical hemostasis is essential to avoid hematoma, which can lead to cellular spread beyond the primary site and biopsy tract. Once the wound is dry and hemostasis is secure after tourniquet release, the