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Epidemiology and Demographics for Primary Vertebral Tumors

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The vertebral column is the most common osseous site for secondary malignancy. Up to 90,000 new cases of spinal metastases occur in the United States annually. Conversely, primary tumors of the vertebral column are relatively rare, with an overall prevalence of 2.5 to 8.5 cases per 100,000 persons per year. They comprise only 10% or less of all tumors to the spine, and total new cases of primary vertebral tumors are estimated at 7500 per year in the United States. The occurrence of lumbar stenosis, cervical radiculopathy, and acute spinal cord injury is estimated at 300 cases, 83 cases, and 5 cases per 100,000 persons per year, respectively.

Population distribution of primary vertebral tumors

The distribution and grade of vertebral tumors varies significantly with age (Table 1). In adults, hemangioma is the most common benign tumor of the spine (20%-30%) [1]. Autopsy studies have shown that up to 10% to 20% of the general population has vertebral hemangiomas, but most are asymptomatic with less than 5% presenting with symptoms [2–4]. Other common benign tumors in adults include osteoblastomas (10%) and osteochondromas (<5%) [5,6].

The most common malignant primary spinal neoplasm in adults is plasmacytoma (30%) [7–9]. Although plasmacytomas are lymphoproliferative tumors, they are included as vertebral tumors because they commonly arise from the bone marrow

* Corresponding author. E-mail address: jchi529@yahoo.com (J.H. Chi). of the vertebral bodies. Other malignant tumors in adults include chondrosarcoma (10%) and osteosarcoma (<5%) [1,10].

In children, the most common benign vertebral tumors are osteoid osteoma/osteoblastoma (12%), aneurismal bone cyst (10%), and eosinophilic granuloma (12% to 25%) [11–13]. Although eosinophilic granuloma is an inflammatory process, it resembles a destructive neoplasm and thus is included here. The most common malignant primary vertebral tumor in children is Ewing's sarcoma (4%–10%) [1,11].

Benign, locally aggressive tumors

Chordoma and giant cell tumor represent a small subset of primary vertebral tumors that are histologically benign but locally aggressive (see Table 1). These tumors make up a unique and distinct group that many experts consider both benign and malignant. Chordoma represents about 2% to 4% of all spinal column tumors and has a high propensity for the sacral and cervical regions [8,14-16]. Although these tumors frequently do not metastasize, they recur locally in an aggressive fashion. Giant cell tumors demonstrate World Health Organization grade I histopathology but also may recur locally. More ominously, these tumors have the potential to metastasize to the lung; therefore many consider these tumors as malignant [17,18].

Gender and racial distributions

Overall, there is a slight male predilection for primary vertebral tumors. Osteoid osteoma,

Table 1
Epidemiology of primary tumors

Tumor	% of Tumors	Cell of origin	Age range	Location	CT/MRI appearance	Growth	Treatment
Benign							
Osteoid osteoma / osteoblastoma	10–12	Osteoblasts	1st and 2nd decades	Lumbar Cervical Posterior elements	Sclerotic, nidus	Slow	Nonsteroidal anti- inflammatory drugs Curettage Radiosurgery?
Chondroma/ osteochondroma	4	Mesenchymal/ cartilage	1st through 4th decades	Cervical Spinous/transverse process	Lytic, circumscribed, non-enhancing	Slow	Resection if symptomatic
Aneurysmal bone cyst	10	Vascular	1st through 3rd decades	Lumbar Posterior elements Vertebral bodies	Cystic/lytic Fluid-fluid level Hemosiderin	Non-neoplastic	Excision Curettage Embolization
Hemangioma	30	Vascular	4th through 6th decades	No preference	Honeycomb Cystic/lytic	Non-neoplastic	Resection if aggressive Embolization Radiation?
Fibrous dysplasia	n/a	Bone marrow stomal cells	1st decade or adulthood	No preference	Lytic, ground glass	Non-neoplastic	Resection if symptomatic
Eosinophilic granuloma Benign, locally aggress	12–25	Histiocytes	1st and 2nd decades	Thoracic Lumbar	Expansile, lytic, vertebrae planae	Non-neoplastic	
Chordoma Chordoma	2–4	Notochord	4th through 6th decades	Sacrum/coccyx (50%) Mobile spine (15%)	Lytic, enhancing, calcification, infiltrating disk space	Slow, but locally aggressive	En bloc Radiation Proton beam
Giant cell	5	Osteoclastic giant cells	2nd and 3rd decades	Sacrum Thoracic Lumbar	Cystic/lytic Nonsclerotic + Hemosiderin enhancing	Slow, but locally aggressive, metastasis to lung	En bloc Embolization Radiation?
Malignant							
Plasmacytoma	20–30	Plasma cells	5th and 6th decades	Thoracic	Lytic "Punched out"		Chemosensitive Radiation sensitive
Chondrosarcoma	7–12	Chondrocytes	3rd through 5th decades	Thoracic	Lytic Destructive "Ring and arc" calcification, enhancing	Malignant	En bloc Resection Chemo/radiotherapy insensitive
Osteosarcoma	< 5	Osteocytes	4th and post-6th decades	No preference	Lytic Destructive Enhancing Osteoblastic	Malignant	Chemo/radiation sensitive Surgery

osteoblastoma, osteochondroma, plasmacytoma, chordoma, and chondrosarcoma all occur more frequently in men than women, in general by a ratio of 2:1 [5,6,16]. Aneurysmal bone cysts and giant cell tumors have a slight female predominance [19,20], whereas hemangioma and osteosarcoma are equal between sexes [4,21]. So far, no ethnic or racial predilection for primary vertebral tumors has been reported.

Anatomic distribution

Certain primary vertebral tumors occur preferentially along the spinal axis and within the vertebrae (see Table 1). Chordomas have a proclivity for the sacrum and coccyx (50%), followed by the skull base (35%) and the mobile spine (15%) [1,22]. Giant cell tumors also tend to occur in the sacrum and thoracolumbar spine. Plasmacytomas and chondrosarcomas are found most commonly in the thoracic spine [10]. Aneurysmal bone cysts and osteoid osteoma/osteoblastomas are found more often in the lumbar spine. Chondroma has a predilection for the cervical spine, especially C2.

Within individual vertebrae, certain tumors have particular preferences for origin. Osteoid osteoma/osteoblastomas and chondromas typically involve the posterior arch including the lamina, pars, and spinous process. Aneurysmal bone cysts also typically involve the posterior arch but also often involve the vertebral bodies. Chordomas usually originate from the vertebral bodies, as do plasmacytomas and giant cell tumors, with varying involvement of the posterior elements.

Signs and symptoms

The most common symptom for primary vertebral tumors is nocturnal back pain. Compression fractures may accompany lytic lesions such as hemangioma, giant cell tumor, eosinophilic granuloma, and plasmacytoma. Neurologic signs, such as myelopathy, may be present with epidural compression. The time course of symptoms also can suggest whether a lesion is benign (slow onset) or malignant (insidious onset) (see Table 1). Pain responsive to nonsteroidal anti-inflammatory drugs is typical of osteoid osteoma.

Diagnosis

When a primary vertebral tumor is suspected, work-up should include pan-imaging including CT and MRI with gadolinium, and angiography,

positron emission tomography and bone scans as indicated (see Table 1). Core biopsy is strongly encouraged to help with appropriate treatment planning. Because of the problem of sampling error and poor tissue yield, needle biopsy has fallen out of favor among leading experts.

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