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Osteoblastoma: A Spectrum of Presentation and Treatment in Pediatric Population

ALEXANDER GOLANT, B.A. AND JOHN P. DORMANS, M.D.

Osteoblastoma is a rare, benign, bone-forming tumor that is histologically related to the more common osteoid osteoma. It is a tumor of the younger population, with nearly 90% of patients diagnosed before age 30 years. Osteoblastoma has been reported in a variety of skeletal locations, but has a distinct predilection for the axial skeleton, particularly the posterior elements of the vertebral column. Despite its benign nature, the tumor may exhibit aggressive behavior and, therefore, typically is treated with surgical excision or curettage. Reconstruction may be required in cases where surgical removal of the tumor creates a sizable defect. Furthermore, osteoblastoma may recur after incomplete removal and, therefore, careful diagnostic evaluation, preoperative planning for adequate and appropriate tumor resection, and a thorough post-operative follow-up are necessary to ensure an optimal outcome. The authors present a review of six children with osteoblastoma, treated at The Children's Hospital of Philadelphia. The authors also review the literature on osteoblastoma, including information about its epidemiology, clinical presentation, radiographic and histologic features, differential diagnosis, natural history, and approach to treatment and follow-up.

The Spectrum of Presentation and Treatment of Pediatric Osteoblastoma

Description of Six Cases Treated at the Children's Hospital of Philadelphia

Patient 1 (AH) was a 12-year-old boy who presented with a 2-month history of worsening left elbow pain. On exam there was an area of bony fullness and tenderness, and also decreased range of motion of the left elbow. Radiographs showed a well-circumscribed $2 \times 2 \times 1.5$ cm mixed area of peripheral reactive sclerosis and central radiolucency involving the distal humerus, with a narrow zone of transition and associated cortical thickening (Figs. 1A, B). A CT scan revealed a nondisplaced pathologic microfracture through the lesion (Fig. 1C). MRI did not indicate a soft tissue mass. Intralesional incisional biopsy with intraoperative frozen sections and subsequent extended curettage and autologous bone grafting were performed. Histologic findings were consistent with osteoblastoma (Fig 2). There was no evidence of tumor recurrence at a 3-year follow-up.

Patient 2 (TH) was a boy who presented to the authors' institution at the age of 5 years, after having been followed by an outside orthopaedist for 3 years for a lesion of his proximal left tibia. At the time of presentation to the Children's Hospital of Philadelphia, the patient's symptoms had worsened to the point where the patient had leg pain daily and was unable to climb stairs. He also had a 15-degree flexion contracture of the knee, 3 cm of thigh atrophy, and a palpable bony fullness of his left proximal tibia. Radiographs revealed a radiolucent diaphyseal lesion surrounded by radiodense sclerotic reactive mature bone (Figs. 3A, B). A CT scan showed marked cortical thickening, but no evidence of soft tissue involvement (Fig. 3C). Incisional biopsy was performed, and after the frozen section confirmed a benign lesion consistent with osteoblastoma, an extended curettage and autologous bone grafting were done. Final histologic examination of the specimen was consistent with osteoblastoma (Fig. 4). Four years after his initial surgery patient had a local recurrence of osteoblastoma; it was treated with extensive intralesional curettage. At a follow-up visit 4 years after his second operation patient had no clinical or radiologic evidence of tumor recurrence.

Patient 3 (DR) was an 11-year-old boy who presented with pain and swelling over his upper sternum. Plain radiographs and a CT scan of his manubrium showed a well-circumscribed expansile radiolucent lesion, surrounded by a thin periosteal shell of bone (Fig. 5). The patient had an open incisional biopsy with intraoperative frozen section, which revealed a benign neoplastic process with osteoid formation. The histologic appearance was consistent with the final diagnosis of osteoblastoma (Fig. 6). Arteriography and embolization of feeding vessels to the tumor were performed and the patient then had resection of the tumor with wide surgical margins. The surgical procedure included reconstruction of the sternal defect with methylmethacrylate and Marlex mesh (CR Bard, Murray Hill, NJ). At a 1-year follow-up the patient was asymptomatic, with full function of his upper extremities, and no evidence of recurrent or residual mass.

From the Division of Orthopaedic Surgery at The Children's Hospital of Philadelphia, Philadelphia, PA.
Address correspondences to: John P. Dormans, M.D., Division of Orthopaedic Surgery, The Children's Hospital of Philadelphia, 34th and Civic Center Blvd., Wood Building, 2nd Floor, Philadelphia, PA 19104.

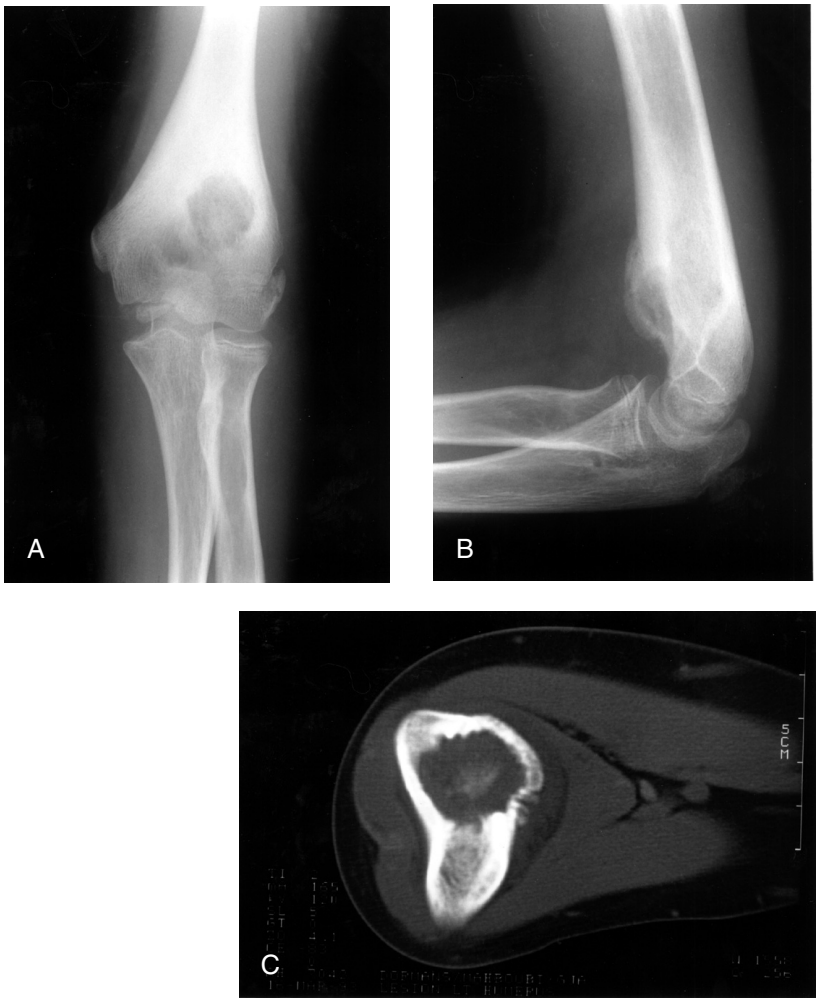

Fig. 1. Patient 1 (AH) – (**A**) AP and (**B**) lateral radiographs of the left elbow demonstrate a $2 \times 2 \times 1.5$ cm tumor in the distal humerus with mixed areas of peripheral sclerosis and central lucency; (**C**) axial CT image of the lesion shows a microfracture through the anterior cortex. Reprinted with permission from Tan, Dormans, Conrad.

Patient 4 (JQ) was a 13-year-old boy who had persistent back pain after a fall that occurred 8 months prior to presentation to the authors' institution. Three weeks prior to admission he developed acute left leg weakness. The patient was transferred to the authors' institution, where plain radiographs showed a process involving formation of new bone in the posterior vertebral elements just below the thoracolumbar junction (Fig. 7A). Bone scan showed increased uptake in the L1 and L2 regions, while MRI and a CT scan showed a large compressive mass with apparent ossification at the approximate level of L1 with significant epidural compression of the thecal sac (Figs. 7B, C, D). An urgent biopsy with subsequent spinal decompression and resection of the tumor was performed with L3-T12 laminectomy, instrumentation and L4-T9 fusion with iliac bone graft. Histologic analysis of the specimen was consistent with osteoblastoma. Postoperatively, patient's lower extremity weakness resolved and within 2 months he was discharged to follow-up with his local orthopaedist. There was no clinical or radiographic evidence of tumor recurrence at a 4-year follow-up.

Patient 5 (JG) was a 3-year-old boy who presented with worsening intermittent left leg pain of 2 months duration. On physical examination the patient was noted to have a limp, moderate left distal thigh swelling, and a slightly decreased range of motion of the left knee. Radiographic studies demonstrated an eccentric, lytic lesion of the distal femoral methaphysis, with extensive surrounding periosteal

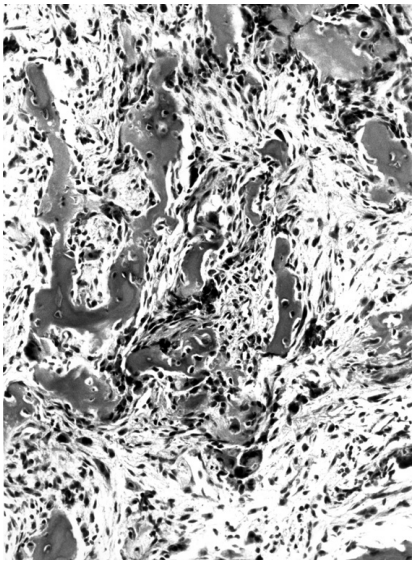

Fig. 2. Patient 1 (AH) – photomicrograph of the lesion demonstrates bony trabeculae with osteoblastic lining. Reprinted with permission from Tan, Conrad, Dormans.

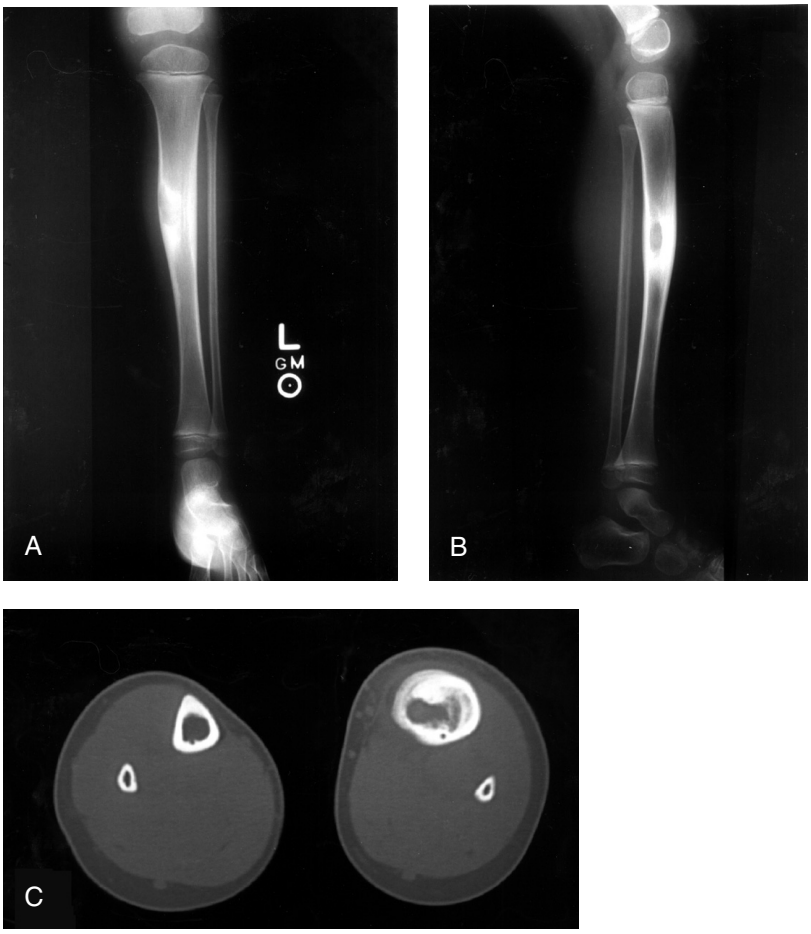

Fig. 3. Patient 2 (TH) – (**A**) AP and (**B**) lateral plain radiographs, and (**C**) axial CT scan of the proximal left tibia demonstrate a diaphysial lesion with marked cortical thickening and a central lucent nidus. Reprinted with permission from Tan, Dormans, Conrad.

bone formation; there was no soft tissue mass but the lesion was associated with extensive edema in the surrounding soft tissues (Fig. 8). Open incisional biopsy was performed with intraoperative frozen sections, and the histologic examination showed an osteoid-producing lesion with prominent osteoblastic rimming in a fibrovascular stroma; there was no histologic evidence of malignancy. The diagnosis of osteoblastoma was made. Extended curettage with bone grafting was then performed. At a 1-year follow-up after his surgery, the child had no clinical or radiographic evidence of tumor recurrence, no pain, normal function and full range of motion of his left lower extremity.

Patient 6 (LY) was a 6-year-old girl who was followed for scoliosis and lipomatosis of paraspinal muscles for 2 years prior to presentation to the author's institution. She had had two prior operations for her lipomatosis. Upon presentation the patient was noted to have fullness and tenderness to palpation in the midline lumbar region, and also an increase in her scoliosis. She was otherwise asymptomatic and had a normal physical exam. Plain radiographs showed thoracolumbar scoliosis (Figs. 9A, B). CT scan and MRI showed a process involving the lower thoracic spine with bony destruction, primarily involving the posterior elements, including the laminae, without extension to the vertebral bodies (Figs. 9C, D). Patient underwent open biopsy with intraoperative frozen sections (consistent with osteoblastoma), and subsequent bilateral decompression and excision of the tumor, and titanium posterior instrumentation and fusion. At a 5-year follow-up, plain radiographs and CT scan of the spine did not show any evidence of tumor recurrence.

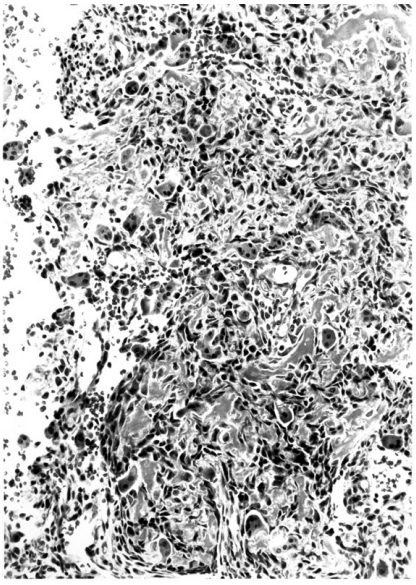

Fig. 4. Patient 2 (TH) – histology of the lesion shows interlacing osteoid trabeculae with surrounding giant cells and prominent vascularity that is characteristic of osteoblastoma. Reprinted with permission from Tan, Dormans, Conrad.

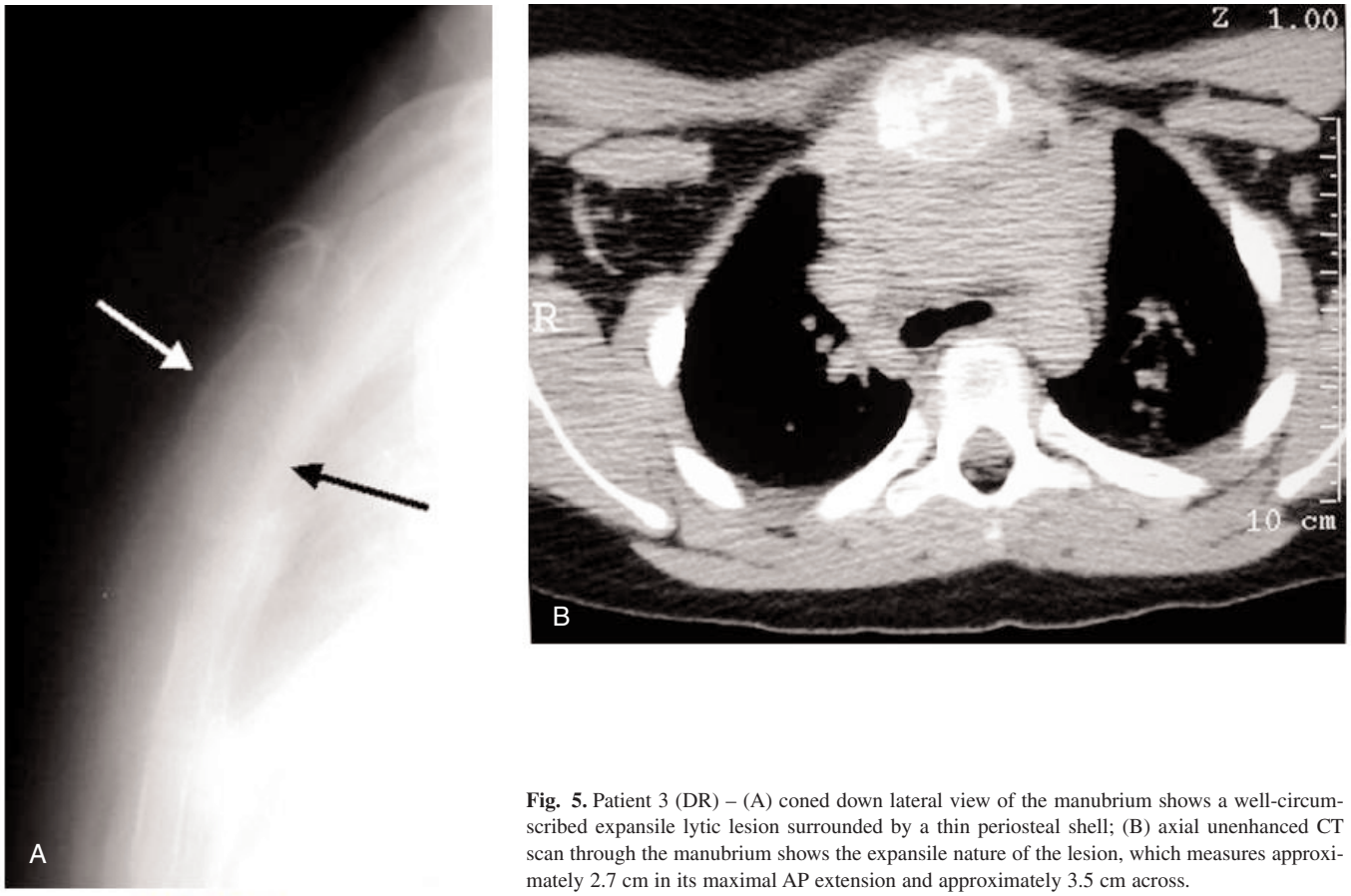

Fig. 5. Patient 3 (DR) – (A) coned down lateral view of the manubrium shows a well-circumscribed expansile lytic lesion surrounded by a thin periosteal shell; (B) axial unenhanced CT scan through the manubrium shows the expansile nature of the lesion, which measures approximately 2.7 cm in its maximal AP extension and approximately 3.5 cm across.

Osteoblastoma – Review of the Literature

Osteoblastoma is a very rare benign bone tumor that is histologically likely related to a more common osteoid osteoma. It was identified as a separate neoplastic entity by Jaffe and Lichtenstein independently in 1956 [11,14,15]. Osteoblastoma accounts for less than 1% of all bone tumors, and approximately 3.5% of all benign bony neoplasms. Males are affected approximately twice as often as females. The average age of incidence of osteoblastoma is between 15 and 20 years, with approximately 90% of patients diagnosed before the age of 30 years [9,26]. While over a 100 cases of benign primary neoplasms of bone are seen annually at the Children's Hospital of Philadelphia only 6 cases of osteoblastoma were seen in the orthopaedic surgery

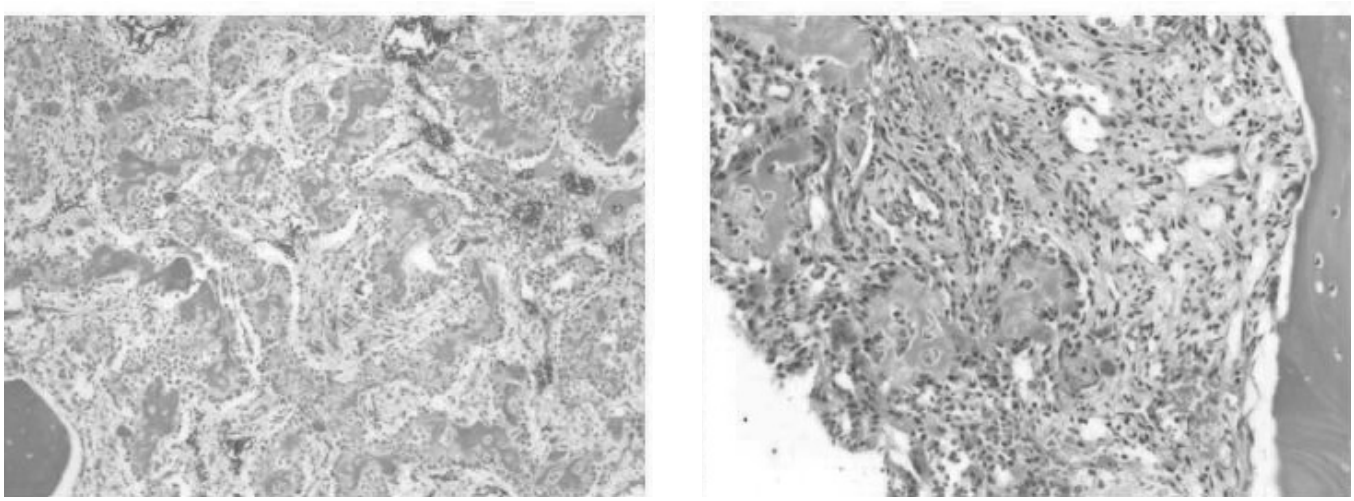

Fig. 6. Patient 3 (DR) - histologic examination at (**A**) 10× and (**B**) 20× magnification shows numerous small irregular osteoid lamellae, rimmed by osteoblasts, set within a vascular stroma, with some osteoclastlike giant cells present. There is a clear interface between the lesion and the host bone.

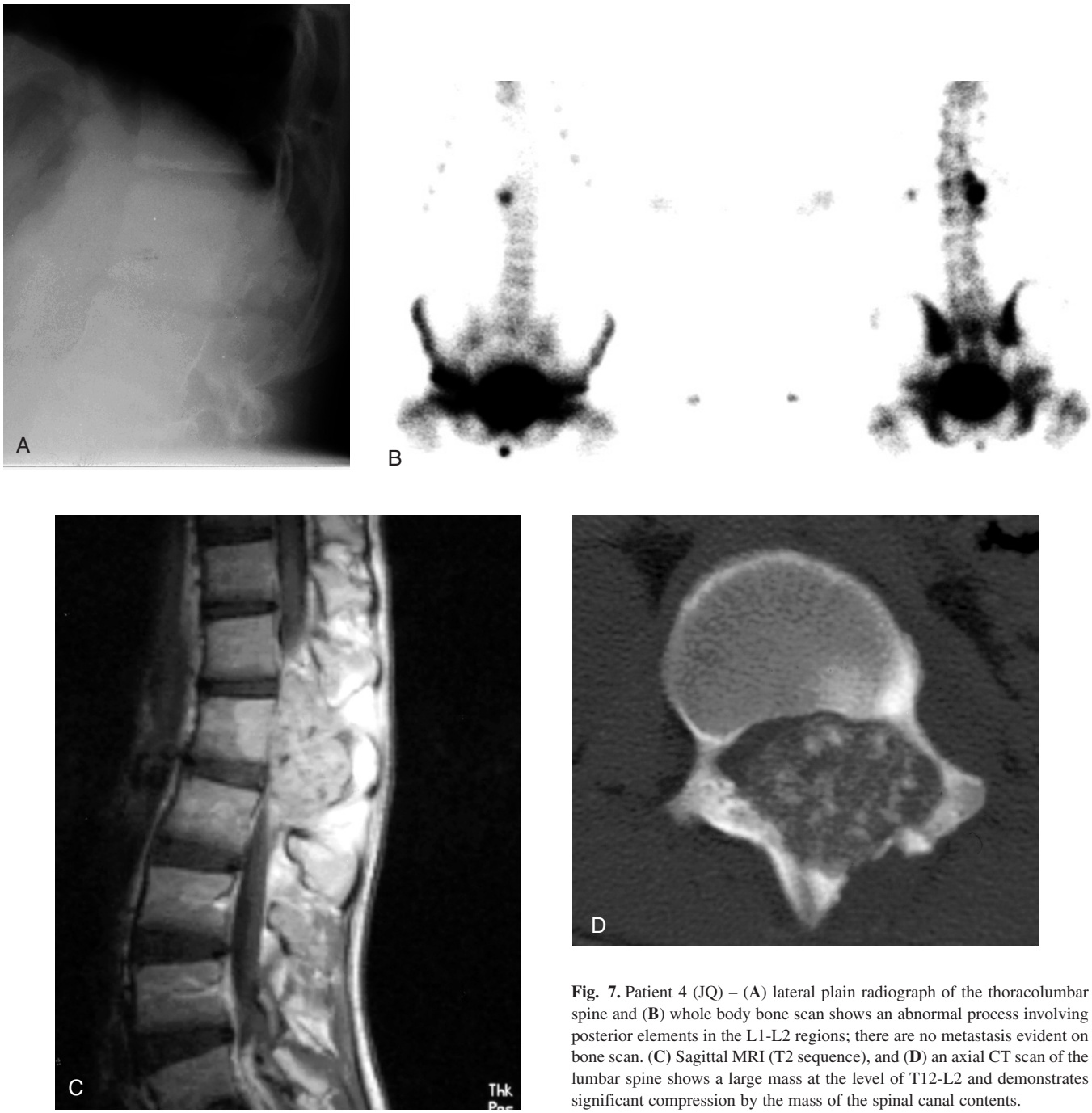

Fig. 7. Patient 4 (JQ) – (**A**) lateral plain radiograph of the thoracolumbar spine and (**B**) whole body bone scan shows an abnormal process involving posterior elements in the L1-L2 regions; there are no metastasis evident on bone scan. (**C**) Sagittal MRI (T2 sequence), and (**D**) an axial CT scan of the lumbar spine shows a large mass at the level of T12-L2 and demonstrates significant compression by the mass of the spinal canal contents.

department over a 10-year period (1999-2002). This underscores the rarity of the osteoblastoma in general and in the pediatric population in particular.

Osteoblastoma has a distinct predilection for the axial skeleton in general and for the posterior elements of the vertebral column in particular. Osteoblastoma also has been reported commonly in the long bones of the appendicular skeleton and the mandible, less frequently in the small bones of the hands and feet, in the skull, maxilla, clavicle, ribs, and rarely in the sternum [9,13,23,26].

The most common presenting symptom of osteoblastoma is insidious onset of pain. The character of pain has been described as dull, aching, often progressive in intensity, and usually localized to the site of the tumor. Unlike the pain of its close relative, osteoid osteoma, the pain of osteoblastoma may not be necessarily nocturnal and is not readily relieved by salycilates [8]. Tenderness at the tumor site is a consistent physical finding, and swelling may be present, particularly when the lesion is near the surface. Patients with spinal tumors may present with paraspinal muscle spasms, scoliosis, neurologic deficits or gait abnormalities [3,7,9,17,26]. Occasionally, osteoblastoma may be asymptomatic and is discovered incidentally [8]. A case of osteoblastoma associated with systemic symptoms has been described in the literature [21], as well as two examples of osteoblastoma associated with osteomalacia [27].

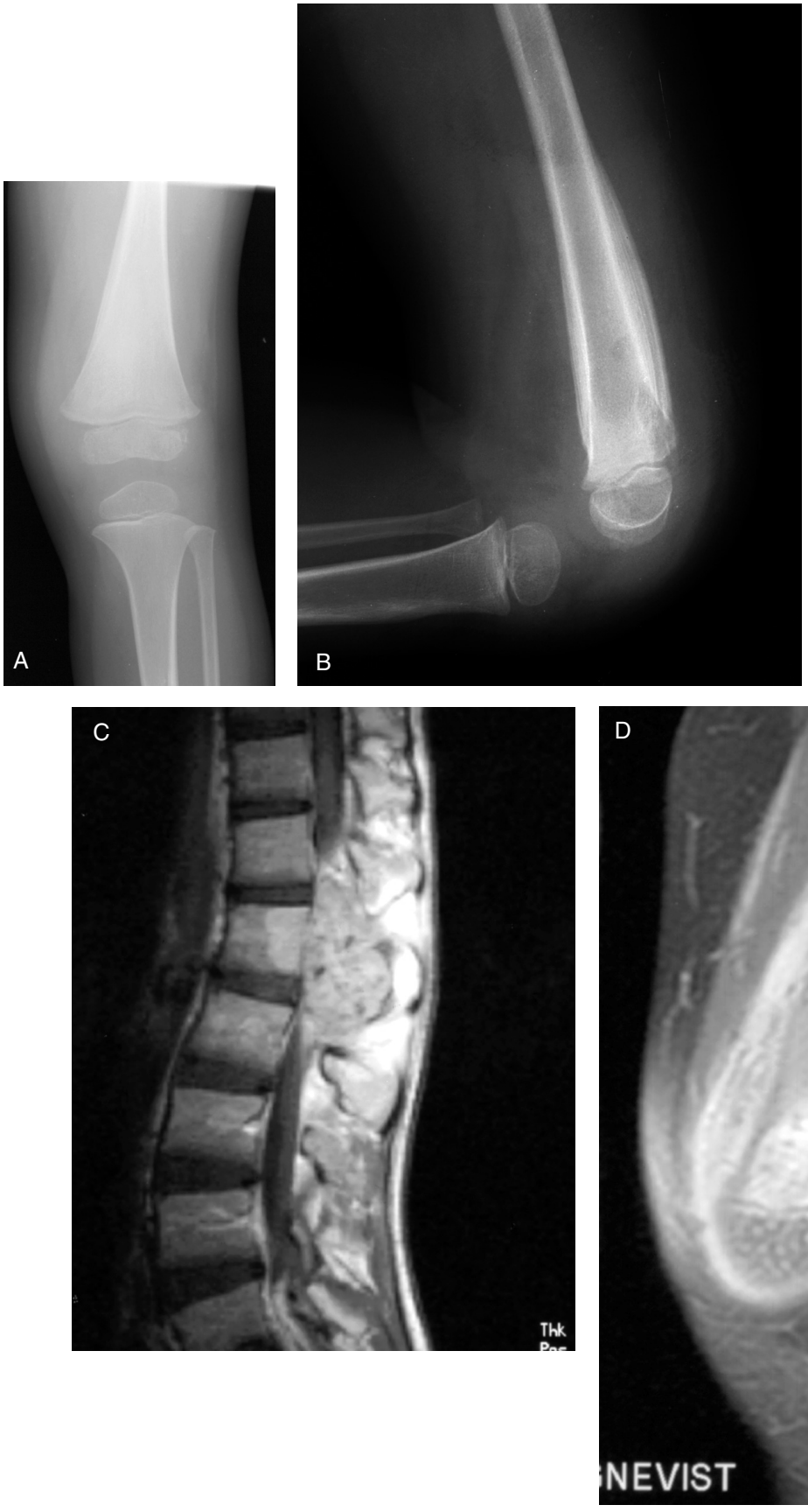

Fig. 8. Patient 5 (JG) – (**A**) AP and (**B**) lateral plain radiographs, (**C**) axial and (**D**) coronal MR images (T2 sequence) of the left distal femur demonstrate an eccentric cortical lesion, extensive reactive sclerosis, and surrounding soft-tissue edema.

Radiologic features of osteoblastomas vary widely and often are nonspecific. In general, they appear as well-circumscribed, rounded or ovoid, central to slightly eccentric areas between 2 and 10 cm in diameter, with an average size of 3 cm [7,18]. The lesions are typically radiolucent (osteolytic) with occasional central radiodensities. The majority

of lesions have no visible central nidus but variable amounts of ossification may be seen in the interior of the tumor. Generally osteoblastomas are expansile lesions, but in majority of cases the cortex remains intact. Although some lesions may exhibit considerable surrounding reactive sclerosis, many only show a thin sclerotic rim. Periosteal new bone formation is common, and usually is benign and of the solid type [8, 18, 20]. The behavior of occasional aggressive lesions, often referred to as "aggressive osteoblastomas" or "malignant osteoblastomas," may simulate that of malignant neoplasms, thereby complicating differentiation of osteoblastoma from osteosarcoma in certain situations. Aside from osteosarcoma, radiographic differential diagnosis of osteoblastoma includes osteoid osteoma, aneurysmal bone cyst, unicameral bone cyst, eosinophilic granuloma (Langerhans cell histiocytosis), chondrosarcoma and enchondroma.

Computed tomography scanning is an important imaging modality for facilitating the diagnosis of osteoblastoma and showing its exact size and location [13]. Intralesional bone production that often does not appear on standard radiographs often is evident on a CT scan, and the bony shell that surrounds the tumor is also shown best on CT scans [7]. In addition, CT scans delineate well areas of cortical destruction and soft tissue extension [8]. Computed tomography scanning is particularly helpful in working up lesions of the axial skeleton [1]. On the radionuclide bone scan osteoblastoma has increased uptake. The bone scan localizes the lesion, but it is not specific enough to plan a surgical resection for lesions in the axial skeleton. Magnetic resonance imaging (MRI) may help to rule out other lesions, which, unlike osteoblastoma, may present with a soft tissue mass. MRI is also useful in evaluating extension and involvement of the epidural space by spinal osteoblastomas. Finally, although it is not commonly ordered, angiography may be used to demonstrate the highly vascular nature of the tumor.

Grossly, the specimen often is hemorrhagic, granular, and friable because of its vascularity and its osteoid component, which shows variable calcification [26]. Histologically, osteoblastomas are well-circumscribed lesions composed of

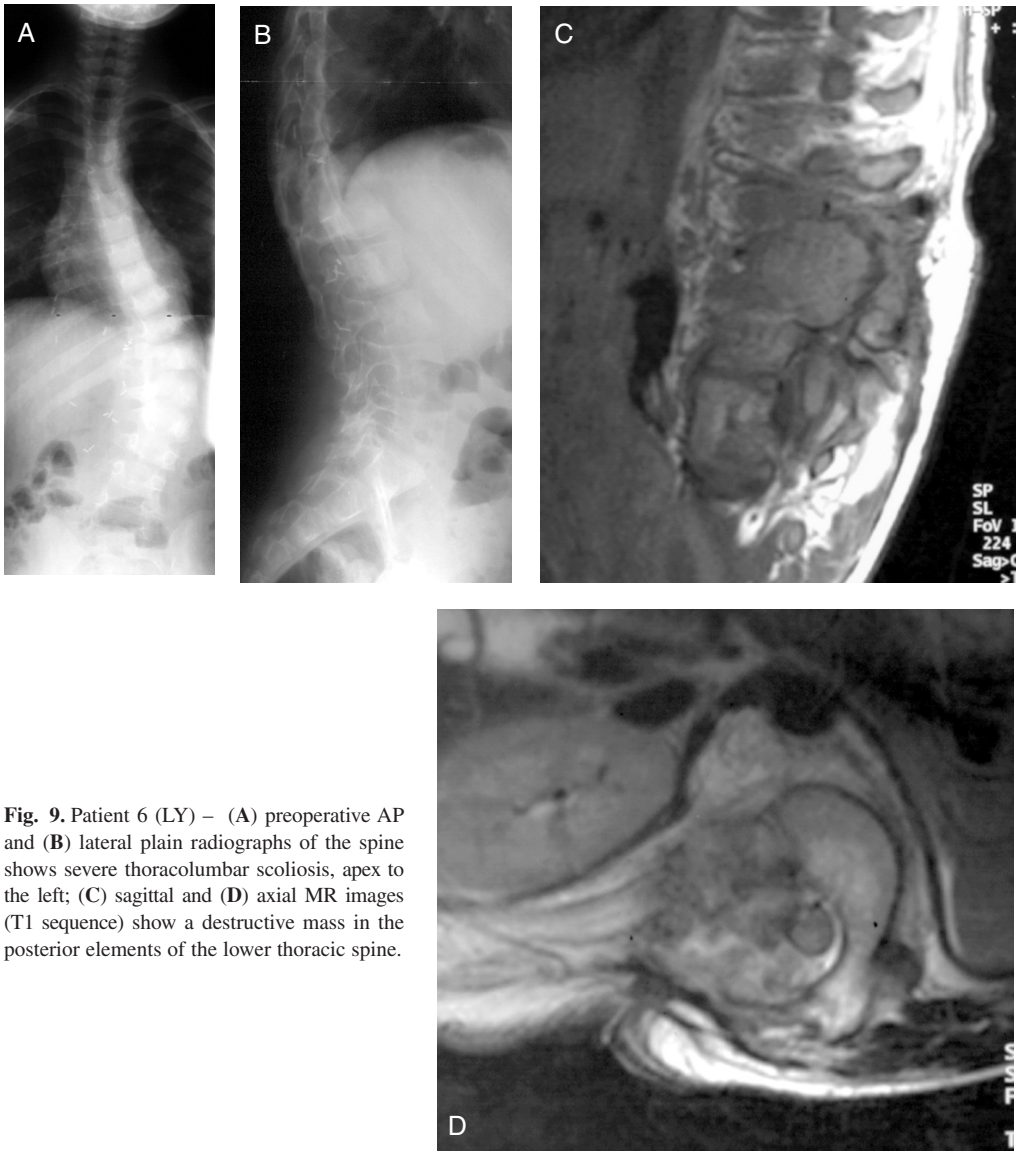

Fig. 9. Patient 6 (LY) – (**A**) preoperative AP and (**B**) lateral plain radiographs of the spine shows severe thoracolumbar scoliosis, apex to the left; (**C**) sagittal and (**D**) axial MR images (T1 sequence) show a destructive mass in the posterior elements of the lower thoracic spine.

immature anastomosing bony trabeculae situated within the abundant fibrovascular stroma. The trabeculae are lined with a single layer of osteoblasts, which are responsible for osteoid or bone formation [9]. The lesions also show variable numbers of osteoclasts and multinucleated giant cells at the surfaces of the bony trabeculae, which are involved in bone resorption.

Osteoblastoma sometimes is called giant osteoid osteoma, underscoring its close histologic resemblance to osteoid osteoma and its larger size [14,18]. However, despite striking histologic similarity between these two lesions, osteoblastoma is considered a distinct clinical entity, differing from osteoid osteoma in clinical presentation and radiologic characteristics [8]. Furthermore, osteoblastoma does not share the limited growth potential of osteoid osteoma and often has a more aggressive clinical course [26]. Distinguishing osteoblastoma from osteoid osteoma can be difficult, both on radiologic and histologic grounds. The most useful differential factor is the size of the lesion: osteoblastoma is a larger lesion measuring 2 cm or more, in contrast to osteoid osteoma which usually is less than 1 cm in diameter. In addition, the clinical manifestations of these two lesions are distinct. Osteoid osteoma commonly is accompanied by nocturnal pain promptly relieved by salicylates, whereas osteoblastoma does not interfere with sleep and does not readily respond to salicylates [3, 8]. The intensity of reactive sclerosis on radiologic evaluation also is helpful, because, unlike osteoid osteoma, osteoblastoma does not consistently exhibit pronounced peripheral sclerosis [9, 15].

Differentiating osteoblastoma from its malignant counterpart, osteogenic sarcoma, may be a difficult task because osteosarcomas resembling osteoblastomas (on radiologic and histologic grounds) and also malignant-appearing osteoblastomas have been reported in the literature [2,4,5,12,18,22]. However, osteosarcoma typically tends to be more invasive; it usually causes cortical destruction, invasion of surrounding tissues, and a more aggressive periosteal reaction. Although osteoblastoma and osteosarcoma can produce osteoid, osteoblastoma does not show the malignant histologic features (cellular anaplasia, atypical mitoses, or permeation of surrounding tissue with entrapment of host bone) seen in osteosarcoma. Also, osteosarcoma exhibits sheets of osteoblasts, while in osteoblastoma only a single layer of these cells lines the bony trabeculae. Another helpful histologic finding for distinguishing osteoblastoma from osteosarcoma is the interface between the tumor and the host bone. In osteoblastoma there is a clear distinction without invasion.

Osteoblastomas are benign, but locally aggressive lesions. Their clinical course may range from slow indolent progression to rapid aggressive growth. Majority of osteoblastomas are active stage 2 lesions and if not treated with surgical resection many may continue to enlarge and damage the bone and adjacent structures [6,25]. According to Enneking, the minority of osteoblastomas are "pseudomalignant" stage 3 lesions, with atypical aggressive clinical, radiographic, and histologic characteristics; these lesions "have a propensity for pervasive recurrence and have been fatal in vital anatomic locations" [6].

Resection with wide margins is preferred for the lesions in expendable bones (rib, fibula), but an extended intralesional curettage is sufficient for lesions in most other locations. As much of the lesion should be removed as possible. Age of the patient must be taken in consideration when planning surgery for osteoblastoma, since limb length discrepancy may result from damage to growth plates in extremities, and spinal deformity may result in cases of vertebral lesions.

Most osteoblastomas are controlled by the extended curettage, but recurrence is not uncommon. A conventional osteoblastoma has a reported recurrence of approximately 10% to 20% [7,10,18]. Neither chemotherapy nor postoperative radiation therapy are indicated [9].

Intricate bony anatomy of the vertebral column complicates the evaluation of and approach to spine tumors. As many as 50% of osteoblastomas are located in the vertebral column, almost always in its posterior elements [1,3,6,18]. Osteoblastomas of the spine require special attention because of the technical difficulty associated with removing the lesion without damaging the spinal cord and related structures. Prompt removal of spinal lesions often prevents or reverses progression of spinal deformity and neurologic deficits. Stability of the vertebral column is an important consideration and should be secured with fusion and instrumentation when necessary. More aggressive procedures are possible in the thoracic and lumbar spine than in the cervical region, but because of the increased axial load vertebral reconstruction and alignment may be more problematic in the lower spinal regions. In all vertebral locations great caution is required to preserve the blood supply of the cord [6].

Finally, as mentioned above, "aggressive" or "malignant" osteoblastomas have been described in the literature on the basis of their unusual clinical behavior and atypical radiologic and pathologic features. These tumors may represent a continuum between typical benign osteoblastomas and low-grade osteoblastic osteosarcomas [5,22], or they may represent the previously misdiagnosed, unrecognized cases of osteosarcoma, since some osteogenic sarcomas may show areas that are histologically indistinguishable from osteoblastoma [2,9]. There are rare reports of spontaneous malignant transformation of osteoblastoma with distant metastases (most commonly to the lungs) [19,24], although it has been suggested that these may represent previously misdiagnosed cases of osteosarcoma. However small (less than 5%), the risk of such transformation and metastasis does exist, and therefore chest x-ray should be obtained in all patients with osteoblastoma (as was done in all osteoblastoma patients seen at the Children's Hospital of Philadelphia).

Osteoblastoma is a benign, but locally aggressive lesion that has been reported in various skeletal locations. Osteoblastoma presents a challenge to the clinician in diagnosis and treatment. The six cases described above illustrate the variable presentation of osteoblastoma and emphasize the need to individualize the treatment approach depending

on patient age, clinical and radiographic findings, and tumor location in the peripheral versus axial skeleton. High quality imaging, careful biopsy providing sufficient material to the pathologist, and meticulous analysis are required to reach the accurate diagnosis. Osteoblastomas should be treated surgically when detected. Close follow-up is warranted for early detection of recurrence, as well as to assure appropriate rehabilitation and recovery.

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