Paget's disease clinical features

Paget's disease has affinity for axial skeleton, long bones and skull. In approximate descending order of frequency: pelvis, thoracic and lumbar spine, skull, femur, tibia, fibula, and clavicles.

Only ≈ 30% of pagetic sites are symptomatic [1)], the rest are discovered incidentally. The overproduction of weak bone may produce bone pain (the most common symptom), predilection for fractures and compressive syndromes: cranial nerve, spinal nerve root... Painless bowing of a long bone may be the first manifestation. A number of patients present due to pain from joint dysfunction related to PD.

The overwhelming majority of pagetic lesions are asymptomatic [2)] with lesions detected on radiographs or bone scan obtained for other reasons or as part of a work-up for an elevated alkaline phosphatase. Although the most common complaint in patients with Paget's disease is of back pain, this is attributable to pagetic involvement alone in only ≈ 12% [3)] in the remainder it is secondary to other factors.

Symptoms that may be related to the Paget's disease itself

Symptoms from the following are slowly progressive (usually present for>12 months; rarely<6 mos):

1. neural compression

a) causes of compression

● due to expansion of woven bone

● due to osteoid tissue

● pagetic extension into ligamentum flavum and epidural fat [4)].

b) sites of compression

● spinal cord

● nerve root in neural foramen

Paget's disease involving the skull: 8th nerve involvement (deafness) is most com- mon. Optic nerve atrophy, and palsies of oculomotor, facial, IX, XI, olfactory nerves and others may also occur [5)].

2. osteoarthritis of facet joints (Paget's disease may precipitate osteoarthritis) Symptoms from the following tend to progress more rapidly:

1. malignant (sarcomatous) change of involved bone (rare)

2. pathologic fracture (pain usually sudden in onset)

3. neurovascular (compromise of vascular supply to nerves or spinal cord) by

a) compression of blood vessels (arterial or venous)

b) pagetic vascular steal (see below)

Paget's disease may present to the neurosurgeon as a result of:

1. back pain: usually not as a direct result of vertebral bone involvement

2. spinal cord and/or nerve root symptoms:

a) compression of the spinal cord or cauda equina (relatively rare)

b) spinal nerve-root compression

c) vascular steal due to reactive vasodilation adjacent to involved areas

3. with skull involvement:

a) compression of cranial nerves as they exit through bony foramina: 8th nerve is most common, producing deafness or ataxia.

b) skull base involvement →basilar invagination.

4. to ascertain diagnosis in unclear bone lesions of the spine or skull.

Paget's disease causes affected bone to weaken, resulting in pain, misshapen bones, fractures and arthritis in the joints near the affected bones. Rarely, it can develop into a primary bone cancer known as Paget's sarcoma. Often Paget's disease is localized to only a few bones in the body. The pelvis, femur, and lower lumbar vertebrae are the most commonly affected bones. Paget's disease typically is localized, affecting just one or a few bones, as opposed to osteoporosis, for example, which usually affects all the bones in the body.

A later phase of the disease is characterized by the replacement of normal bone marrow with highly vascular fibrous tissue.

Spinal cord symptoms

Myelopathy or cauda equina syndrome may be due to spinal cord compression or from vascular effects (occlusion, or “steal” due to reactive vasodilatation of nearby blood vessels [6] Only ≈ 100 cases had been described as of 1981 [7] Characteristically, 3–5 adjacent vertebrae are involved [8] , whereas monostotic involvement is usually asymptomatic [9].

In case reports in the literature, progressive quadri- or paraparesis was the most common presentation [10]. Sensory changes are usually the first manifestation, progressing to weakness and sphincter disturbance. Pain was the only symptom in a neurologically intact patient in only 5.5%.

A rapid course (averaging 6 wks) with a sudden increase in pain is more suggestive of malignant degeneration.

The clinical triad of gait disturbances, memory impairment and urinary incontinence is associated with a communicating hydrocephalus in the 'normal pressure hydrocephalus'-syndrome. The authors present a case of H.D.C. (hydrocephalus-dementia-complex) in Paget's disease with an identical syndrome, but with obstructive hydrocephalus, causing a triventricular dilatation. Today, each case of mental deterioration in Paget's disease, should be immediately observed and neurosurgical intervention kept in view. An X-ray of the skull, a CT-scan of the brain and a cisternography are performed as routine procedure. If there are indications of involvement of the basis of the skull, hydrocephalus and/or disturbed pattern of the tracer-migration around the convexities, associated with a certain degree of dementia, impaired gait or urinary incontinence, a ventriculo-subcutaneous drain should be inserted. If clinical improvement follows, a ventriculatrial shunt is indicated. The post-operative clinical outcome seems to be dependent on some clinical and technical factors. The most eventful outcome is observed in cases where the clinial triad, described in patients with a classical 'normal pressure hydrocephalus'-syndrome, is associated with an obstructive hydrocephalus, due to a stenosis of the Sylvian aqueduct [11].

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