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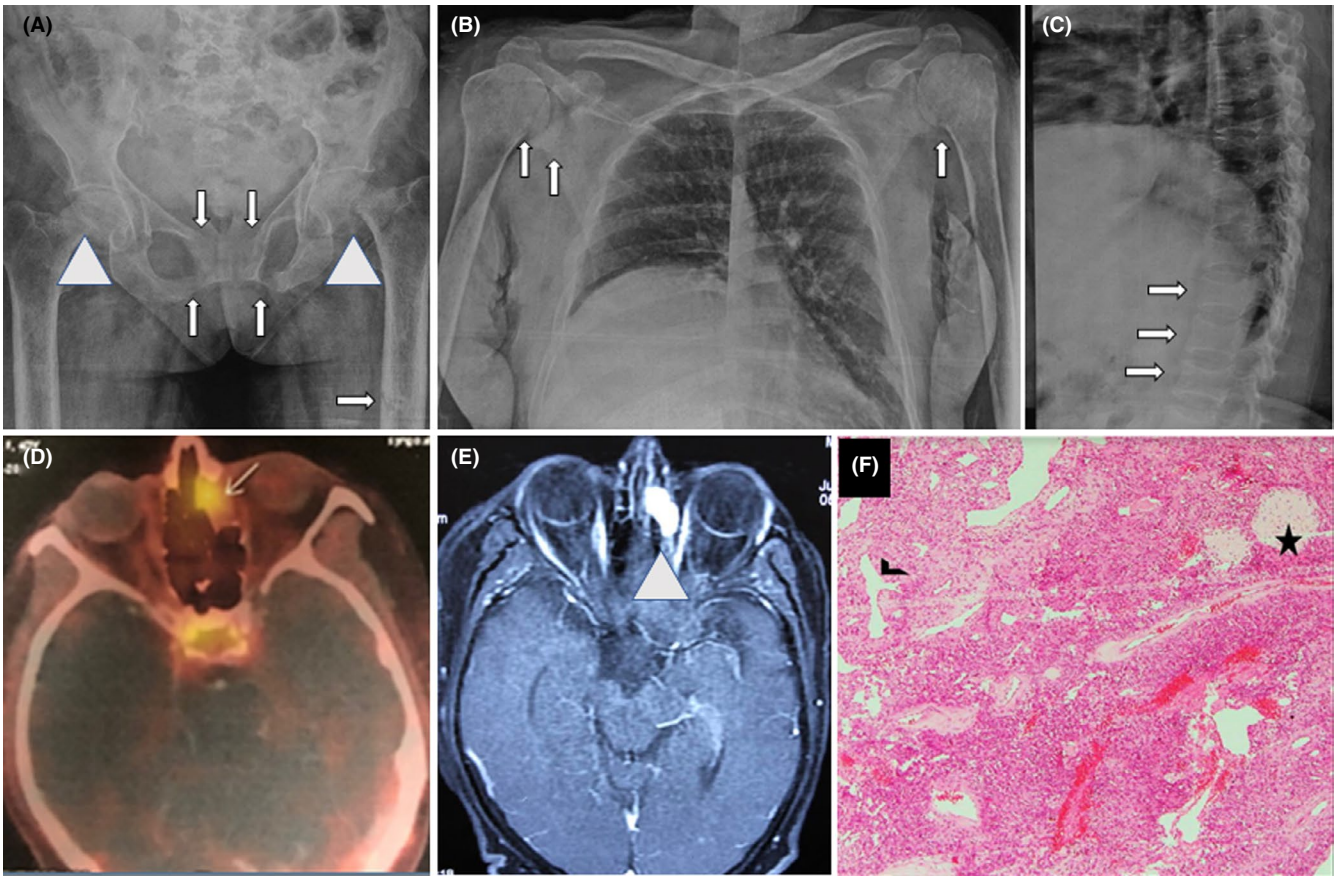
## Clinical Image: Multiple pathological fractures in a middle-aged woman: A rheumatologist's challenge




The patient, a 40-year-old woman, presented with diffuse muscle pain and cramps with progressive difficulty in walking for the last 4 years. She had thoracic kyphosis and tenderness over both legs and the proximal shoulder and thigh regions. **A and B**, X-rays revealed multiple bilateral fractures involving the neck of the femur (arrowheads) and the shaft of the fibula as well as the looser zones in the pubic rami, the shaft of the femur, the scapula, and the humerus (arrows). **C**, She had multiple codfish vertebrae (arrows). Ionic calcium and 25-hydroxy vitamin $D_3$ levels were normal. The fasting serum phosphate level was 1.9 mg/dl (normal: 3-5 mg/dl), the alkaline phosphatase level was 1367 IU/l (normal: less than 240 IU/l), and the serum parathormone level was 85 pg/ml (normal: 15-65 pg/ml). The bone mineral density revealed severe osteoporosis (*z* score at spine: –4.4). Further investigations revealed reduced tubular reabsorption of phosphate and reduced tubular maximum reabsorption of phosphate as well as its higher fractional excretion (44.5%; normal: less than 5%), suggestive of renal phosphaturia and possible tumor-induced osteomalacia (TIO). **D**, She was advised to undergo a $^{68}$Ga-DOTANOC positron emission tomography/computed tomography (PET/CT) scan, which showed increased uptake at the left nasal bone near the nasion (arrow). **E**, T1-weighted contrast magnetic resonance imaging (MRI) of the brain with facial cuts showed a well-defined, lobulated, heterogeneously enhancing mass lesion in the left cribriform plate of ethmoid epicentred at nasion (arrowhead). Fibroblast growth factor 23 (FGF23) and 1,25-dihydroxyvitamin $D_3$ (1,25$[OH]_2D_3$) levels could not be estimated. She was initiated on 1 g/day of sodium phosphate (uptitrated to 3 g/day) in divided doses with calcitriol 0.25 µg/day. Despite medical therapy, the phosphate level did not normalize, and she was advised to undergo surgical resection of the tumor. Post successful resection, her phosphate levels became near normal, with gradual fracture healing, enabling walking with minimal support at 6 months' follow-up. **F**, The histopathology revealed cellular tumor showing branching blood vessels (arrowhead) and monomorphic ovoid cells with a focus of cartilaginous metaplasia (asterix) suggestive of hemangiopericytoma (Hematoxylin & Eosin, magnification 200x) , the most commonly encountered phosphaturic mesenchymal tumor. A diagnosis of TIO is often delayed because of nonspecific musculoskeletal complaints and its rarity. The characteristic biochemical abnormalities

include hypophosphatemia, normal or low $1,25(OH)_2D_3$ levels and normal or elevated FGF23 levels (1). TIO is typically caused by benign, slow-growing mesenchymal tumors originating either from bones or soft tissues. The most common location of these tumors is the thigh and femur (22.7%), followed by the craniofacial region (20.7%) (2). TIO is commonly associated with hemangiopericytomas, although hemangiomas, sarcomas, and fibromas have also been reported (1). Functional imaging with octreotide single-photon emission computed tomography, DOTANOC PET/CT, or fluorodeoxyglucose–PET/CT is the preferred investigation to locate the tumor, followed by anatomic localization with computed tomography and/or MRI (3). Surgical resection is definitive, but medical management is needed in the interim or when resection is not possible because location or multiplicity.

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