Staging studies for evaluation of squamous cell carcinoma

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Answer: Staging of left temporal squamous cell carcinoma with PET-CT.

Diagnosis

Primary hyperoxaluria with a focal oxalomatous lesion mimicking metastatic disease.

Discussion

Primary hyperoxaluria is a rare congenital disorder of abnormal glyoxylate metabolism with resultant over-accumulation of oxalate, which then binds with calcium and precipitates in the kidneys, resulting in nephrocalcinosis and nephrourolithiasis. As renal function worsens, patients develop systemic oxalosis with calcium oxalate deposition in the soft tissues, musculoskeletal system, cardiovascular system, central nervous system, and eyes [1–5].

There are three common types of primary hyperoxaluria (types I–III). Type I is the most common and severe and

involves a mutation in the alanine glyoxylate aminotransferase (AGT) enzyme causing overproduction of oxalate glyoxylate [1]. Type II results from deficiency of glyoxylate reductase and hydroxypyruvate reductase (GRHPR), which results in overproduction of oxalate and L-glycemic aciduria [1, 5]. Type III results from mutation in 4-hydroxy-2-oxoglutarate aldolase (HOGA) and is the least aggressive hyperoxaluria type, with most patients not progressing to end-stage renal disease (ESRD) [1].

Imaging findings in primary hyperoxaluria vary depending on patient age and renal function [2–6]. Prior to renal function impairment, imaging is frequently normal. As renal failure develops, cortical nephrocalcinosis and nephrourolithiasis will be seen (Fig. 1). Calcium oxalate subsequently deposits in soft tissues, tendons, and joint capsules, with joint effusions and nodular-appearing tumoral calcinosis (especially in periarticular regions, which can cause erosive arthropathy) [2–6]. As ESRD develops toward the late stage of the disease, musculoskeletal imaging manifestations typically associated with renal osteodystrophy and secondary hyperparathyroidism will develop, such as osteosclerosis, subperiosteal resorption, “rugger-jersey” spine, and vascular/soft tissue calcifications (also seen in Fig. 1). Abnormal skeletal maturation and pathologic fractures with delayed healing can also be seen but are similarly nonspecific. Oxalate deposition in the subcutaneous and dermal tissues can cause skin necrosis/ulceration, and oxalate deposition in the myocardium, pericardium, ocular and vascular regions has also been described [4–9]. Skeletal findings more specific to primary hyperoxaluria may include irregular diaphyseal transverse sclerotic bands and wide metaphyseal zones [5].

Lytic lesions can be seen in primary hyperoxaluria, which histopathologically represent foci of calcium oxalate deposition with a granulomatous foreign body histiocytic reaction, which appear essentially identical to brown tumors on

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Fig. 1 AP abdominal radiograph (left); coronal CT image of the abdomen/pelvis on bone window (right)

imaging (Fig. 2, upper left) [6]. Information about the activity of such a lesion on 18-fludeoxyglucose (FDG) PET imaging is scarce in the literature, but our case shows that such a lesion can be hypermetabolic (Fig. 2), highlighting that these lytic lesions should be interpreted with caution in hyperoxaluria patients with a history of cancer.

Following biopsy of a lytic lesion, histopathologic examination with H&E staining will show irregular trabeculae of predominantly woven bone (usually characteristic of hyperparathyroidism) with marked crystal deposition between trabeculae (Fig. 3, upper right and upper left). Arrangements of crystals in “flower” and “star-shaped” forms are characteristic

Fig. 2 Axial CT (upper left) on bone window, axial corrected PET (upper right), axial fused PET-CT on bone window (lower left), and coronal fused PET-CT on bone window (lower right)

Fig. 3 Histopathologic images using hematoxylin and eosin (H&E) stains at low power (20×, upper left), high power (40×, upper right), and high power polarized microscopy (40×, bottom)

of calcium oxalate, and nearby granulomatous foreign body histiocytic response is frequently seen. Polarized microscopy will also demonstrate extensive crystal deposition, some with needle-like extension (Fig. 3, bottom) [1, 5, 6]. Needle-like crystals can also be seen with gout, which can also coexist with calcium oxalate deposits; therefore, gout should be excluded.

Treatment of primary hyperoxaluria is generally supportive and involves hydration, increasing urine pH (for nephrourolithiasis prevention), and pain control [1, 5]. Renal and liver transplantation can significantly improve clinical outcomes, and dialysis is a viable option while awaiting transplantation, although patients are still at higher risk of mortality [1].

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Conflicts of interest The authors declare that they have no conflicts of interest.

Disclosures None.

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