Raised CA19.9 and hepatic space occupying lesion after teriparatide therapy in a case of polyostotic fibrous dysplasia

Sir,

We have previously submitted case report in our journal titled "Spinal polyostotic fibrous dysplasia in two adults: Does only biopsy unravel the mystery?"[1] Case-2 was treated with bisphosphonates and narcotic analgesics without any relief and became bed bound. PFD is a disease of admixture of mutated cells with abnormal cAMP levels and normal cells. Over the time bony lesions in PFD get normalized.[2] It is hypothesized that mutated cells undergo apoptosis following period of rapid turnover like puberty and proliferation of remaining normal cells leads to normalization. Parathyroid hormone (PTH) is stimulant for osteoblastic cells, and may lead to early apoptosis of mutated cells. Our patient had low bone mineral density at lumbar spine with compression fractures. Hence after

taking ethical clearance and written consent from patient, we started patient on teriparatide therapy 20 μg subcutaneously daily. After two months, patient reported for review. His clinical condition had deteriorated. Re-evaluation revealed deterioration with fresh lesion appearing on PET-CT [Figures 1 and 2]. Space occupying hypoechoic lesions appeared in liver on ultrasonography, CT scan and FDG PET (SUV - 6.3). New lesions were also seen in bones with maximum SUV at anterior superior iliac spine (5.7). Oncological markers showed elevated CA19.9 (671 U/ml normal < 37). In view of above malignancy was suspected. FNAC of liver showed normal liver cells. Open biopsy from anterior superior iliac spine demonstrated classical Chinese letter pattern [Figure 3] again confirming the diagnosis of PFD.

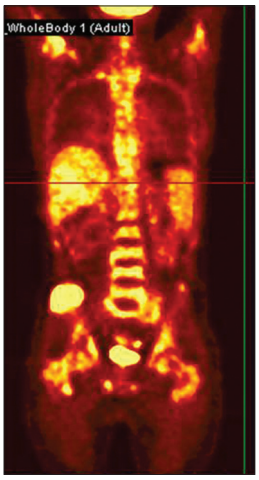

Figure 1: FDG-PET showing avid uptake in spine, humerus, liver, and pelvis

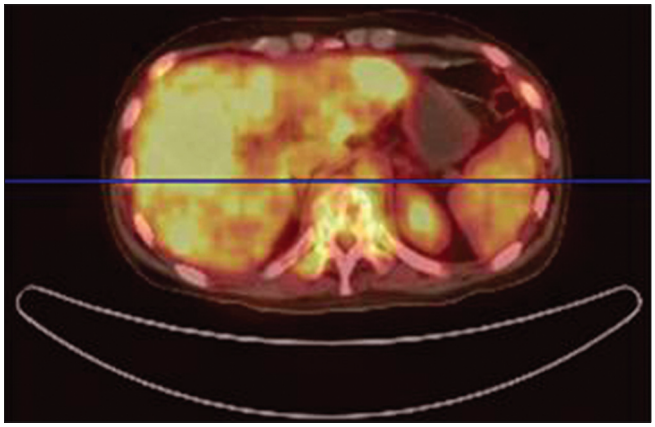

Figure 2: Multiple FDG avid space occupying lesions in liver

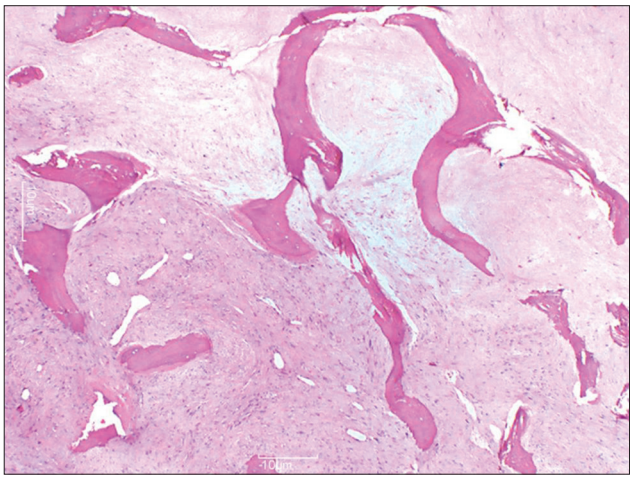

Figure 3: H and E, ×10 staining showing classical Chinese letter pattern in bone biopsy

To best of our knowledge, this is the first report of raised CA19.9 levels in PFD. CA 19.9 (carbohydrate antigen 19.9), also known as sialylated Lewis a-antigen (a blood protein in red blood cells), is an antigen defined by the monoclonal antibody 1116NS 19.9. It was first mentioned by Koprowski *et al*.[3] CA19.9 is synthesized by varied cells in the body like normal human pancreatic and biliary ducts and by gastric, colonic, endometrial, salivary and bronchial epithelium. False positive values are reported pancreatobiliary disorders, inflammatory bowel disease, hypothyroidism, kidney problems in up to 31%.[4] After thorough search we could only find one case report where an intramuscular myxoma was associated with elevated CA 19.9 levels.[5] Intramuscular myxomas are associated with GNAS1 mutation and their association with PFD is known as Mazabraud's syndrome.[6] Our patient also developed hepatic space occupying lesion and FNAC revealed normal hepatic morphology suggestive of hepatic nodular hyperplasia. Elevation of CA19.9 has also been reported with liver disorder. Our patient had persistently elevated liver enzyme for last one year without evidence of viral infection or autoimmunity. Deterioration in our patient can be either due to teriparatide therapy or some unknown environmental agent. If it was due to PTH associated stimulation of Gsα receptor, it can be speculated that other drugs or chemicals stimulating Gsα receptor can also cause deterioration and may require to be avoided.

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