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Fibrous Dysplasia In Elderly Patient With Painfull Hips

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Özet

Fibröz displazi (FD) kemik maturasyonunda bozuklukla giden, çok fazla fibröz dokunun üretilerek trabeküler kemik dokusu yapımında zayıflıkla giden bir kemik hastalığıdır.

Algoloji kliniğimize neredeyse 10 yıldan beri olan kronik kalça ağrısı nedeni ile başvuran bir olguyu sunuyoruz. Ağrısı oturmakla artıyormuş. Ağrı nedeni ile 5-10 dk'dan daha fazla yürüyemiyormuş. Yatarak istirahat etmekle ağrısı azalırken NSAID ve diğer analjeziklerle ağrısı kısmen rahatlıyormuş. 2 yıl öncesinde spondilolistezis nedeni ile opere edilen hastanın ağrı şikayetinde bir azalma olmamış. Hasta aynı zamanda papiller tiroid kanseri nedeni ile tiroidektomize imiş. Pelvis ve kalça ekleminin direk grafisinde sağ sakroiliak eklemde ve ramus ossis pubiste skleroz görülmekteydi. Lomber MRG ile sağ sakroiliak eklemde osteitis kondensans ilei olup ramus ossis pubis üzerinde 7,5x 4 cm ebatlarında etrafı sklerotik kemikle çevrili iyi sınırlı litik expansil kitle izlenmekteydi. Lezyon symphisis pubise doğru uzanmakta olup öncelikli olarak fibröz displaziyi düşündürüyordu. Kemik lezyonu daha sonra PET-BT ile değerlendirilmiş olup L1-L2 vertebralar ile sol 7-8. Kostaların lateral kenarlarında artmış aktivite tutulumu vardı. Hastanın eşlik eden malignitesi olması nedeni ile metastaz şüphesini dışlamak için biyopsi ile doku tanısına gidilmiştir. Biyopsi ile FD tanısı ne doğrulanabilmiş ne de ekarte edilebilmiş olup lezyonda malignite dışlanabilmiştir. FD olgularının tanı ve takiplerinde PET-CT oldukça güvenilir ve non-invaziv bir yöntem olup hastalar mutlaka PET-CT görüntüleri ile değerlendirilmelidir. FD kür edilebilen bir hastalık değildir. Hasta cerrahi eksizyon için ortopediye konsulde edilmiş olup ileri yaşı nedeni ile cerrahiye uygun bulunmamıştır. NSAID ve opiatlardan oluşan palyatif tedavisi düzenlenerek hasta malign transformasyon açısından takibe alınmıştır.

Klinik deneyimlerimizde kalça ağrısı ile başvuran yaşlı bir hastada fibröz displazi oldukça nadirdir. Bu hastalarda malign transformasyon olabileceği için tanı ve takiplerinde PET-BT kullanılması gerekliliğine vurgu yapılmıştır.

Anahtar Kelimeler: Fibröz displazi, yaşlı hasta

Abstract

Fibrous dysplasia (FD) is a skeletal disorder in which bone-forming cells fail to mature and produce too much fibrous tissue and poorly formed trabecular bone.

We are presenting an elderly woman who refer to our algology clinic with painful hips. Her complaints are in chronic state and have been neerly 10 years. Especially while sitting her hip pain aggrevates. She feels comfortable only bed resting. NSAID and other analgezic drugs slightly alleviates her pain. She hashave history of papiller thyroid cancer. In the overview of pelvis and axial hip joint image there was sclerozis in the right sacroiliac joint and ramus ossis pubis. Lomber MRI was showed that there was a lythic and expansil lesion, 7,5x4 cm in size, on the right ramus ossis pubis and was encovered by sclerotic fine borded bone tissue. Lesion was reaching through the syphysis pubis and thougt firstly FD. This bone lesion invastigated furtherly with bone sythgraphy and seen increased activty on the vertebras L1, L2 and left 7th, 8th costas lateral sides. Furthermore investigation made by PET-CT. There was fracture on the arcus of right 7th Costae and 12th torokal vertebrae. There was hypermethabolic sclerotic bone lesions on the among right ramus ossis pubis, left 8th arcus costae and 7th left costae's anterior side. We exclude any methastatic lesion or malign transformation with bone biopsy. FD is not curable disease. So we gave palyative therapy such as nsaıds and opioids. We recommend follow up for malign transformation.

We report this case to keep in mind that FD could refers with hip pain in elderly patient. As our clinical experience FD is a quite suprising diagnose in elderly patient with hip pain. We also emphasised that nuclear imaging such as PET-CT is quite usefull in diagnose and follow up in bone lesions.

Keywords: fibrous dysplasia, elderly patient

Background

Fibrous dysplasia (FD) is a skeletal disorder in which bone-forming cells fail to mature and produce too much fibrous tissue and poorly formed trabecular bone. In generally FD diagnosed in childrens or young adults. Although patients with FD are commonly asymptomatic it could be present with pain or bone deformities. Bone lesions have characteristic appearance on x rays, which is usually sufficient to make the diagnosis. The diferantial diagnose of lytic lesions in metaphysis or diaphysis with "ground glass" appearences are nonossifying fibrome, unicameral bone cyts, aneurysmal bone cyts, chondromixoid fybrome. Further imagining techniches like as MRI, bone scanes, PET-CT can be helpfull for diagnose, classification and follow up for malign transformation (1). We are presenting an elderly woman patient with painful hips who undiagnosed throughout her life. She has history of papiller thyroid ca and we emphasise importance of nucleer imaging for diferantial diagnose of her pelvic lesions as unexpecting hematogenous bone methastase or any other bone lesions.

Case

78 year old woman refer to our algology clinic because of her painfull hips and backs. Her complaints are in chronic state and have been neerly 10 years. Even her

dorsalgia has been since the age of sixteen, when she had trauma (car crash) to the back. Especially while sitting her hip pain aggrevates and she couldn't walk or stand longly except 5-10 minutes. She feels comfortable only bed resting. NSAID and other analgezic drugs slightly alleviates her pain. She was operated 2 years ago just because of spondylolistezis and her complaints wasn't relieve after surgery.

She was also operated for papiller thyroid cancer. She was given radioactive ıyode therapy 2 times with an interval of 6 months.

By the physical examination she was seen in the flexion posture. She had a deformity of pectus carinatum seen by inspectively. Her lomber paravertebral muscles were painfull and spinose processes were tender. Nervetension tests were negative also sacroiliac compression was painless. Range of motion of hip joint was limited especially while internal and exteral rotation. Both FABER and FADIR tests were limited and produced pain on hip joint. Symphysis pubis was tender while palpating. Lomber extension also restricted. Shober test was measured 3cm. Her neurological examination was normal except hypoactive deep tendon reflexes.

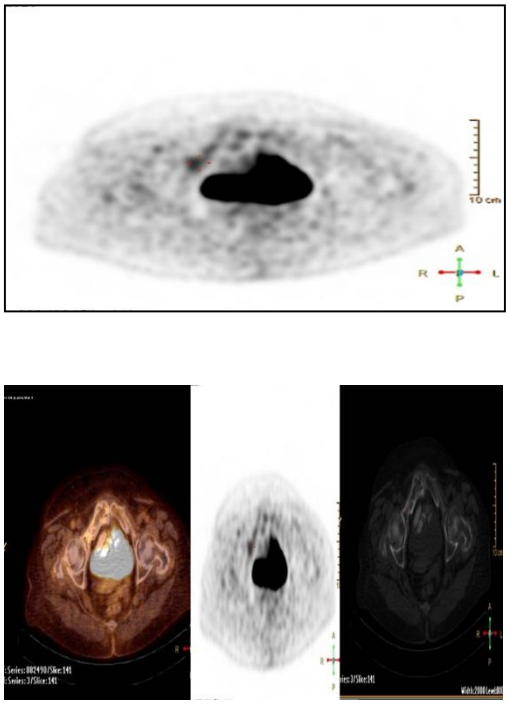
In the overview of pelvis and axial hip joint image there was sclerozis in the right sacroiliac joint and ramus ossis pubis. Lomber MRI was showed that there was osteitis condensans ilei on the right sacroiliac joint and there was a lythic and expansil lesion, 7,5x4cm in size, on the right ramus ossis pubis and was encovered by sclerotic fine borded bone tissue. Lesion was reaching through the syphysis pubis and thougt firstly FD. This bone lesion invastigated furtherly with bone sythgraphy and seen increased activty on the vertebras L1, L2 and left 7th, 8th costas lateral sides. Patient's t score on femur neck was 2.4. Because of the trauma history lesions could be sequelly even could be methastatic whiches primer papiller thyroid ca. Furthermore investigation made by PET-CT. There was fracture on the arcus of right 7th Costae (SUVmax: 6.2) and 12th torokal vertebrae (SUVmax: 4.4). There was hypermethabolic sclerotic bone lesions on the among right ramus ossis pubis (SUVmax: 3.5) (figure-1), left 8th arcus costae (SUVmax: 4.0) and 7th left costae's anterior side (SUVmax: 4.1). to determine the essential diagnose we underwent the patient surgery and bone biopsy was taken. Pathology of the lesion exclude the methastase or malign transformation. But it was not a satisfactoryly material to either diagnosed or exclude fibrouse dysplazia. Patient refused to repeat the biopsy. So we decided with both imagining and the patients clinic that our essential diagnose was fibrous dysplazia.

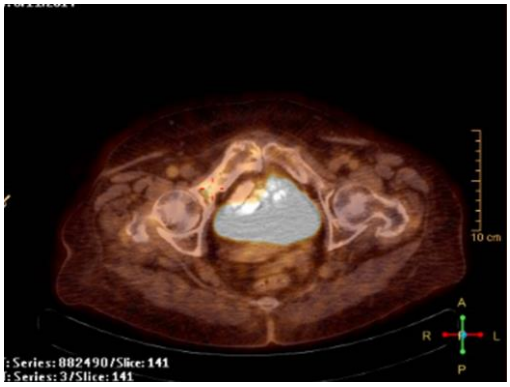
Discussion

In generally FD diagnosed in childrens or young adults. The process originates in the medullary cavity. It is caused by a postzygotic mutation in the guanine nucleotide stimulatory protein (GNAS1) gene. It is more of a skeletal dysplasia than a true neoplasm. This rare disease firstly described by lichtenstein in 1938 (2). It is usually diagnosed in children and young adults, and is present throughout life. If patients have only one bone involved it is called monostotic, whereas more than one bone involvement classified as a poliostatic forms of fibrous dysplazia. It could also be the part of sydromes like as mccune albright sydromes which is associated with endocrine abnormalities and café-au-lait spots or mazabroud sydrome known with soft tissue mixomas and renal missing of phosphat (3, 4).

In elder patients diagnose of disease depends mostly on imaging (5-7). Because of the lack of skin lesions, precocs puberty and endochrinologic pathology diagnostic algorythm based on a probability. To this end guideline recommendations followed and the lesion which previusly been demonstrated by CT confirmed by MRG (5). MRG showed us that patient's lesions are well matched with FD poliostatic lesions (8). We continued algorythm by sythntigraphy for another involvement. One of the lesions on the 8th costae was probably FD, but another one on the 7th costae thought secondary to fracture. Thyroid-CA is the confusing factor in patient's history. PET-CT confirmed that there was a hipermetabolizm on these lesions. F-18 FDG PET/CT can be considered in the management and follow-up of this pathology. FDG avidity on Fibrous Dysplasia can be variable and specific discrimination between Fibrous dysplasia and malignant bone tumors is not well defined, with large SUVmax overlap values (9). Clinical use of PET-CT on benign tumors or tumor like lesions is quite common (10). F-18 FDG PET/CT can also be useful for pretherapeutic assessment, for example in guiding biopsy with a better accuracy for histologic grading or for evaluation of effectiveness of neoadjuvant chemotherapy (11). We exclude any methastatic lesion or malign transformation with bone biopsy. İn order to bone biopsy neither defined nor exclude FD, the essential diagnose made by both clinically and imaging. Other conditions investigated, which well known associates with FD (3,4). If these poliostatic FD lesions were part of syndromes such as mccune albright there must be other conditions for diagnosis. So we learned from patient that she had her very first menstrual siclus when she was abouth 9 or 10 year old. Precocious puberty is usually defined as the onset of secondary sexual development before the age of eight years in girls and nine years in boys (12). In most populations, attainment of pubertal milestones is normally distributed, with a standard deviation of approximately one year, and the mean age of onset of puberty is about 10.5 years of age in girls and 11.5 years in boys. So age of 9 could be acceptable such as a puberte precocious (12-14). Of course we could not know retrospectively either the exact menarsial age of patient because of her eldery or the precense of puberty precocious. We also consultated patient dermataology for cafe au lait and endocrinology for any endochrine pathologies. There wasn't any cafe au lait lesions on the dermatological examination. Also there wasn't any endochrinologic pathology except thyroid function tests. Patient was othyroid but TSH was supressed as seen as usually after surgery while using thyroid hormon replacement. Thyroglobulin was <0.20 ng/ml and whole body pertecnetat scan and the thyroid sythigraphy showed there wasn't any residual or

recurrens or abnormalties. We haven't seen any soft tissue myxomas whith either phsycal examination or imagination techniches. Also urine analise was clear about renal missing of phosphat. So we exclude both the sydromes which mostly accompained with FD mccune albriht sydrome and mazabroud sydromes. Probably our patient have had these lesions since her childhood and haven't been diagnosed until now. Patient was suffering from hip pain and we depend it the pubik bone involvement. FD is not curable disease. There was several study in literatur about the surgical treatment of FD on the proksimal femur (15,16). We could reached only one surgical study which requires 29 patients and also two of them with FD on pubic ramus. One of them treated by curetation and bone grafting and the other one curateted and demineralized bone matrix alone. İn follow up two of FD patient relieved from their hip pain at night (17). But in our case both patient and the surgeons refused the surgery by the several reasons such as elderly and comorbidity. So we gave palyative therapy such as nsaıds and opioids.





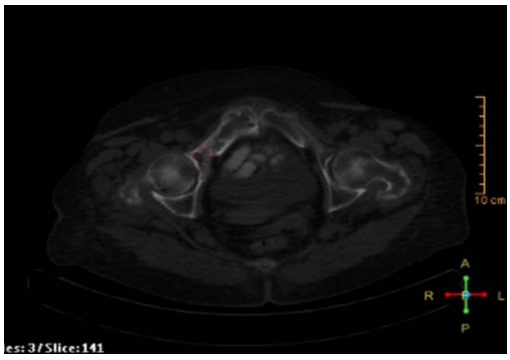


Fig.1. PET/CT scanning was performed after the injection of 222 MBq (6mCi) F-18 FDG. This study demonstrateted hypermetabolic activity in the right ramus ossis pubis.

We recommend follow up for malign transformation. We report this case to keep in mind that FD could refers with hip pain in elderly patient. As our clinical experience FD is a quite suprising diagnose in elderly patient with hip pain. We also emphasised that nuclear imaging such as PET-CT and bone syntigraphy can be helpfull to determine malign transformation and bone methastases; and recommends for follow up for patient with benign bone tumors or tumor like lesions.

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