IOSR Journal of Dental and Medical Sciences (IOSR-JDMS)
e-ISSN: 2279-0853, p-ISSN: 2279-0861.Volume 21, Issue 1 Ser.13 (January. 2022), PP 60-61
www.iosrjournals.org

A Rare Presentation of Takayasu Arteritis

Dr SUBIN S[1] Dr SREEJITH VIJAYAKUMAR[2] Dr SANDRA PRABHU[3]
Dr GEETHA P[4]

[1]*Junior Resident,Department of General Medicine,Govt Medical College,Calicut*
[2]*Medical Officer,Sree Gokulam Medical Centre,Attingal,Trivandrum*
[3] *Medical Officer,Sree Gokulam Medical Centre,Attingal,Trivandrum*
[4]*Professor,Department of General Medicine,Govt Medical College,Calicut*

Date of Submission: 08-01-2022 Date of Acceptance: 23-01-2022

I. Objective

To describe an interesting case of Pyoderma Gangrenosum in a patient with Takayasu Arteritis

II. Introduction

Takayasu arteritis(ICD-10:M31.4) is a rare chronic large vessel vasculitis which predominantly affects aorta and its major branches, otherwise known as pulseless disease seen mainly seen in female of Asian origin less than 40 years of age. It is known for its variable clinical presentation. Takayasu arteritis presenting with pyoderma gangrenosum is extremely rare , described in less than 1 % of all cases. Here we describe the presentation of 22 year old male who presented to the General Medicine OPD for evaluation of Hypertension with a background history of recurrent history of non-healing ulcers in his right thigh for 6 years and after clinical evaluation and relevant radiological investigation ,was diagnosed with Takayasu arteritis.

III. Methods

Clinical history, physical examination, biochemical lab data, imaging and pathology were analysed

IV. Case Presentation

22 year old male with a past history of recurrent history of non-healing ulcer in right medial thigh for 6 years now presented with flare up of ulcer in right thigh for 1 week patient. He was detected to have hypertension and attended Medicine OPD for evaluation of hypertension .

On examination

He was conscious oriented, moderately built and poorly nourished

Ulcer of about 6X6 cm present in medial part of right thigh

Vitals: Pulse Rate-88 per min ;Radiofemoral delay present

BP: upper limb :160/90 right and 150/90 left Lower limb :140/80 right and 140/80 left

CVS:systolic bruit was heard over left carotid artery, subclavian artery and right carotid artery and subclavian artery. Other system examinations were within normal limits

Lab values- HB-14g/dl(normal:12-15g/dl), WBC 6600cells/cubic millimetre(normal 4500-11000 cells/cubic millimetre) , RBC 4.03 million/mm^3 (normal 4.3-5.9 million/mm^3), Hematocrit 44.5%(normal 41%-53%) Platelets 3.51L(normal 1.5-4L), ESR 16mm/hr(normal-20mm/hr) Serum Na 140, SerumK 4, Serum Calcium 9.8, Serum Phosphate 2 ,Blood Urea Nitrogen 24, Serum Creatinine 0.8, Serum Uric Acid 3, Serum Magnesium 1.8, Random Blood Sugar 106, Total Bilirubin/Direct Bilirubin 0.8/0.6, Total Protein/Serum Albumin 6.9/4, Alkaline phosphatase 60, SGOT/SGPT 27/24, TSH 1.64, 24 hr metanephrine negative, UrineRoutineExamination normal

Skin biopsy of ulcer was consistent with pyoderma gangrenosum and was evaluated for its common associations

OGDscopy and biopsy of bowel revealed a non specific enteritis

USG KUB with Renal Artery doppler – kidney – Normal, no evidence of renal artery stenosis

MR AORTOGRAM: Diffuse circumferential wall thickening involving thoracic and abdominal aorta,right and left common carotid artery and subclavian arteries suggestive of Takayasu arteritis

MANAGEMENT

Treated with oral steroid prednisolone 60mg OD for 1 month ,calcium supplementation and ante hypertensive Telmisartan 40 mg HS;supportive management for ulcer given. After 1 week wound started healing and is symptomatically better. Under follow up in OPD

Now BP controlled and tapered dose of steroids

V. Discussion

Takayasu arteritis is a chronic large vessel vasculitis which predominantly affects aorta and its branches. It is seen in young Asian females of less than 40 years of age, diagnosed with ACR criteria. Clinical presentation is heterogenous; can present with constitutional symptoms, hypertension, and cardiovascular features with bruit being the most common clinical sign. It can also present with neurological features like TIA/stroke. Dermatological manifestations are rare with erythema nodosum most common presentation. Pyoderma gangrenosum is an extremly rare association of takayasu arteritis.

VI. Conclusion

Pyoderma gangrenosum in a patient with Takayasu arteritis can be a debilitating condition that is often misdiagnosed due to its rarity. Although rare, physicians must always consider the possibility of such a disease in cases presenting with recurrent non healing ulcer.

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Dr SUBIN S, et. al. "A Rare Presentation of Takayasu Arteritis." *IOSR Journal of Dental and Medical Sciences (IOSR-JDMS)*, 21(01), 2022, pp. 60-61.