



PIGMENTED VILLONODULAR TENOSYNOVITIS: A DIAGNOSIS OF RARITY, OR IS IT????

Clinical Research

Dr Ashwani Umat	Associate Professor, Deptt Orthopaedics Maharishi Markandeshwar Medical College, Mullana, Ambala, Haryana, India
Dr Manjeet Singh*	Head Of Department, Dept. Of Orthopaedics M.M Medical College, Mullana, Ambala, Haryana, India *Corresponding Author
Dr Vishesh Verma	Post Graduate Resident, Orthopaedics Maharishi Markandeshwar Medical College, Mullana, Ambala, Haryana, India
Dr Sonia Kochhar	Head Of Department, Deptt Of Physiology, Maharishi Markandeshwar Medical College, Mullana, Ambala, Haryana, India
Dr Chetan Sharma	Post Graduate Resident, Orthopaedics, Maharishi Markandeshwar Medical College, Mullana, Ambala, Haryana, India
Dr Harsimarjit Kaur	Associate Professor, Deptt Anatomy, Government Medical College And Hospital, Patiala, Punjab, India

ABSTRACT

Background: Pigmented Villonodular tenosynovitis is a benign lesion that is considered by many as a differential of rarity but presents as any regular chronic joint swelling of whom diagnosis and treatment are on totally different lines.

Materials and Methods: The study was conducted on 14 patients who presented with chronic knee or ankle swellings. Complete history and previous management lines were noted and there diagnosis was then mulled with a broader scope of investigations.

Result: Three out of these 14 cases came out to be biopsy proven Pigmented Villonodular Tenosynovitis (PVNS), of which two were around the knee and one was over the ankle. Out of the three, one patient had been on antitubercular regimen for over three months without any relief, one was on DMARDS based upon clinical suspicion while the third one was being treated non-specifically with NSAIDS.

Conclusion: We thus concluded that PVNS must be kept as a viable diagnosis while dealing with chronic knee and ankle swellings.

KEYWORDS

PVNS, DMARDS, NSAIDS, Knee And Ankle Swellings.

Introduction

Pigmented Villonodular Tenosynovitis is a rare and benign condition but is locally aggressive & potentially recurrent.^{1,2} The term was coined by Jaffe, et al. Its localised form was first explained by Chassaignac (1852) who described it as "CANCER OF TENDON SHEATH".^{3,4}

Its characterized by synovial proliferation and hemosiderin deposition inside joints, tendon sheaths, and bursae.⁵

Theories of Origin are multiple but aetiology remains unknown.⁴

Radiographs are mostly unremarkable, but Magnetic Resonance Imaging (MRI) may be characteristic.

The lesion has no specific clinical symptomatology.⁶ Treatment requires complete synovectomy. But even after all efforts for complete removal, recurrence is high.⁷

Patients And Methods

This is a retrospective study conducted over 14 patients who attended orthopaedic Out Patient Department (OPD) and Emergency for chronic knee and ankle swelling unresolving with conventional methods. Patient group was chosen from period between Jan 2016 to 2018. This work is based upon Pre-Operative and Post-Operative blood investigations, clinical, Radiographical, as well as histological data available from patient's records. Age-wise distribution with location of the lesion is depicted in table 1.

(Table-1: Age wise Distribution of Lesion With location)

S. No	Age/ Sex	Location of lesion
1	32 / F	Knee
2	52 / M	Knee
3	48 / M	Knee
4	35 / M	Ankle
5	21 / F	Ankle
6	23 / M	Medial malleolus
7	50 / M	Knee
8	42 / F	Ankle

9	32 / F	Knee
10	44 / M	Knee
11	28 / F	Ankle
12	30 / F	Knee
13	38 / F	Knee
14	25 / M	Knee

Result

Out of 14 patients three patients came out to be biopsy proven PVNS, five patients as Mono Articular rheumatoid Arthritis and six patients as Tuberculosis. Results are depicted in table 2.

(Table-2: Comparing Pre-biopsy and Post Biopsy Diagnosis)

Serial Number	Diagnosis and Treatment	
	Pre Biopsy	Post Biopsy
1	Tubercular on ATT	PVNS
2	TB on ATT	TB
3	TB on ATT	TB
4	Undiagnosed on NSAIDS	RA
5	Undiagnosed on NSAIDS	TB
6	Undiagnosed on NSAIDS	PVNS
7	Undiagnosed on NSAIDS	RA
8	Undiagnosed on NSAIDS	RA
9	RA on DMARDS	RA
10	Undiagnosed on NSAIDS	TB
11	Undiagnosed on NSAIDS	TB
12	RA on NSAIDS	RA
13	RA positive on Mtx and Hcq	PVNS
14	RA on NSAIDS	RA

Discussion

The term PVNS was used by Jaffe to describe idiopathic villous overgrowth of synovial membrane and typical pigmentation (chocolate brown colour as shown in image 3) usually found in a single joint.³ Geschickter and Copeland states that lesion develops due to proliferation of osteoclasts in sesamoid bones.³ The lesion has two primary forms, Localised and Generalised. Localised form occurs predominantly in hands followed by feet, ankle, wrist and elbow,

whereas generalised form occurs most commonly in knee followed by ankle and foot.⁴ Histologically it shows bland fibrous tissue containing histiocytes & giant cells.⁸ Microscopically two types of villi are present (in diffuse form) including one with Coarse “Shag Carpet” appearance and a Fine “Fernlike” villi.⁴ Histologically it shows Bland fibrous tissue containing histiocytes & giant cells.⁸ Radiographs are often unremarkable (Image 1), whereas Magnetic Resonance Imaging (MRI) may show characteristic intra-articular masses with signal dropout on T2-weighted sequences (Image 2).^{9,10} The hypo-intensity is due to interaction of iron in ferric state with water molecules.¹ Clinically the lesion may or may not cause Mechanical symptoms.⁸ But If left untreated it gradually eats up the affected area and surroundings rendering the patient with severe morbidities. Treatment is complete excision of synovium (Image 3,4,5) with or without radiotherapy. Synovectomy with Total joint replacements⁸ offer better pain relief along with lower recurrence rates. Role of Radiotherapy is unclear due to limited data available. Intra Articular radiation synovectomy using Yttrium-90 and dysprosium-165 are still experimental.³

As is evident the diagnosis and treatment of the condition is way different then conventional disorders (like RA and TB) and needs an observant meticulous approach.

It was realised during course of the study that every non- resolving swelling isn't always RA or TB. It was found that 50 percent of total patients in the study group were being treated without a diagnosis with non-specific NSAIDS. In a setup like our country where patients are poor, illiterate and go to quacks for treatment who put them on long term pain killers & steroids we as clinicians should broaden our scope of differentials and PVNS is worthy of being in that list. Clinicians were found initiating ATT and DMARDS based upon clinical suspicion and blood investigations which are less- specific (as in RA factor and raised ESR). In cases of chronic knee and ankle swellings that don't resolve with conventional treatment, MRI and biopsy must be done despite blood investigations and clinical findings suggesting otherwise, especially in non-resolving cases. PVNS may be a diagnosis of rarity but should be kept at the back of the mind while dealing with chronic unresolving swellings.



Image-1: X-Ray Image Of PVNS



Image-2: T1 V/ST2 weighed MRI Images



Image-3: Intra Operative Image Of The Lesion showing characteristic Chocolate Brown Color



Image-4: Intra-Operative Image Showing the Extent Of the Lesion

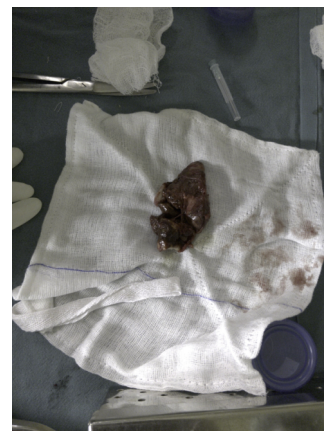


Image-5: Post Excision Image Of P.V.N.S

Conclusion

It was thus concluded that any chronic knee or ankle swelling must be evaluated with a wider scope of thinking with MRI and Biopsy as integral part of the workup to provide better patient care and avoiding adverse drug effects.

References

- 1) M Lee, S Mahroof, J Pringle, S C Short, T W R Briggs, S R Cannon. Diffuse pigmented villonodular synovitis of the foot and ankle treated with surgery and radiotherapy. *International Orthopaedics (SICOT)* (2005) 29: 403–05.
- 2) Stevenson JD, Jaiswal A, Gregory JJ, Mangham DC, Cribb G, Cool P.(2013). Diffuse pigmented villonodular synovitis (diffuse-type giant cell tumour) of the foot and ankle. *Bone Joint J* 95-B(3):384-90.
- 3) Weinstein S, Buckwalter J, Turek S. *Turek's orthopaedics*. 7th ed. 2016.
- 4) Kulkarni G. *Textbook of orthopedics and trauma* (4 volumes). 3rd ed. 2016.
- 5) Chirag Kapoor, Maulik Jhaveri, Rishit Soni, Malkesh Shah, Parth Rathi, Paresh Golwala. Pigmented Villonodular Synovitis of the Knee Joint: A Case Report. *Cureus* (2016) 8(10): e816.
- 6) Margad O, Boukhris J, Azriouil O, Daoudi M, Mortaji A and Koulali K. (2017). Les synovites villonodulaires du genou: à propos de 20 cas. *Pan African Medical Journal*, 28.
- 7) Sung KS, Ko KR.(2015).Surgical outcomes after excision of pigmented villonodular synovitis localized to the ankle and hindfoot without adjuvant therapy. *J Foot Ankle Surg*. 54(2):160-3.
- 8) Azar F, Beaty J, Canale S, Campbell W. *Campbell's operative orthopaedics*. 13th ed. 2017.
- 9) Hao DP, Zhang JZ, Xu WJ, Wang ZC, Wang XN.(2017). Pigmented villonodular synovitis of the ankle: radiologic characteristics. *J Am Podiatr Med Assoc*. 101(3):252-8.
- 10) Kramer DE, Frassica FJ, Frassica DA, Cosgarea AJ.(2009). Pigmented villonodular synovitis of the knee: diagnosis and treatment. *J Knee Surg*. 22(3):243-54.