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Original Article

Polyarteritis Nodosa in Children: A Single-Centre Experience of 23 Patients

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ABSTRACT

Introduction: Childhood Poly-Arteritis Nodosa (cPAN), characterized by systemic necrotizing vasculitis affecting medium- and small-sized arteries, represents approximately 9% of all childhood vasculitis cases. While registries for cPAN existed in other countries, such information was not readily accessible in India. Consequently, this study sought to provide a comprehensive overview of the clinical presentation, findings from histopathological, angiographic, and genetic investigations, as well as the pharmacotherapy employed in the management of cPAN.

Methods: We conducted a retrospective analysis of the electronic medical records of patients with a diagnosis of cPAN, as per the EULAR, PRES, and PRINTO guidelines, who had visited out institute over a period of 13 years (2010 – 2022). Clinical, laboratory, radiological characteristics along with the pharmacotherapy and outcomes were summarized and compared to previously published studies.

Results: A total on N=23 patients' data were analysed. The most common clinical manifestations at diagnosis were fever (n = 22/23; 93%), skin manifestations, (n = 17/23; 74%) myalgia and arthralgia (n = 13/23; 56% each). Glucocorticoids were used in all patients along with a non-glucocorticoid immunosuppressant like Azathioprine, Cyclophosphamide and Mycophenolate Mofetil. The relapse rate was 78.26 % (n=18/23) and mortality was seen in 8.7% (n=2/23)

Conclusions: cPAN is a rare disease of insidious onset with variable clinical presentation and is associated with severe complications despite appropriate treatment. Therefore, early diagnosis and aggressive treatment forms the cornerstone of the treatment.

Keywords: *Immunosuppressants, Outcomes, Paediatrics, Rheumatology, Vasculitis.*

INTRODUCTION

Polyarteritis Nodosa (PAN) is an uncommon type of systemic necrotizing vasculitis primarily impacting medium-sized arteries, with occasional involvement of small arteries. The first documented account of this condition traces back to the mid-19th century, as reported by Adolph Kussmaul and Rudolph Maier. [1] Childhood PAN (cPAN) accounts for nearly 9% of all childhood vasculitis cases. [2] The disease can range from a more benign cutaneous variety to a severe systemic variety more often affecting the musculoskeletal, dermatologic, neurological, gastrointestinal and renal systems. [3] The usual presentation includes fever, arthralgia/arthritis, tender cutaneous nodules, myalgia, weight loss, and hypertension. The affected children also typically have anaemia, leucocytosis, thrombocytosis and elevated acute phase reactants. [4] Histopathology reveals fibrinoid necrosis of the vessel wall that begins in the media but almost always involves the entire internal elastic lamina. [3] Computed tomography (CT) angiography that has replaced conventional angiography in the diagnosis of cPAN show characteristic features such as aneurysms, segmental narrowing, occlusions and variations in the

calibre of arteries, together with pruning of the peripheral vascular tree. [3] Until recently, the aetiology was unknown when in 2014, Cat Eye Syndrome Chromosome Region Candidate -1 (CECR1) mutation leading to deficiency of adenosine deaminase 2 (DADA 2) was shown to be associated with childhood PAN. [5]

Lee et al., [6] Eleftheriou et al., [7] and Sönmez et al., [8] have reported their experience of treating cPAN which have broadened our understanding of this rare disease. However, there is still a paucity of information about cPAN from the Indian subcontinent. Thus, to our best knowledge, the current study is one of the largest series of cPAN from the Indian subcontinent so far with an attempt to bridge the knowledge gap about cPAN. The primary objective of this study was to summarize the clinical presentation; the results of histopathological, angiographic, and genetic investigations and pharmacotherapy in children afflicted with cPAN.

The secondary objective was to compare the findings in the current cohort to previous key publications from around the world.

METHODS:

Ethics:

The institutional ethics committee, Manipal Hospital, Bengaluru approved the study vide letter no…. dated….. A waiver of consent was obtained as the study was retrospective in nature based on medical records with no direct patient/ participant interaction. The research adhered to the ethical principles of Good Clinical Practice that abides by the standards outlined in the Declaration of Helsinki (World Medical Association, Fortaleza, 2013) and followed the National Guidelines for Ethical Research in Human Participants (Indian Council of Medical Research Guidelines, 2017).

Study Design and Setting:

We conducted a retrospective analysis of the electronic medical records (eMR) of patients with a diagnosis of cPAN, visiting the paediatric rheumatology clinic of a tertiary care teaching hospital located in Bengaluru, South India between the years 2010 and 2022.

Eligibility Criteria:

All children of any sex aged less than 18 years of age who satisfied the diagnostic criteria for cPAN as outlined by EULAR, PRES, and PRINTO classification guidelines were included in the study. [3] The diagnostic requirements for cPAN include the confirmation of necrotizing vasculitis in medium or small arteries through histopathological findings or the detection of an angiographic anomaly (such as an aneurysm, stenosis, or occlusion), which is a compulsory criterion. Furthermore, at least one of the following five characteristics must be observed: skin manifestations, myalgia or tenderness in muscles, hypertension, peripheral neuropathy, or renal complications [3] There were no exclusions.

Study Variables and Quantitative Data:

The demographic, clinical, and laboratory features recorded at the time of diagnosis included the following factors: gender, age, ethnicity (determined through information given by patients or parents during hospital registration), affected organs, erythrocyte sedimentation rate (ESR), serum C-reactive protein (CRP) levels, platelet count, the presence of proteinuria and/or hematuria, positivity for antinuclear antibody (ANA) and antineutrophil cytoplasmic antibody (ANCA) (either perinuclear or cytoplasmic), histopathological results, arteriographic findings, pharmacotherapy, and treatment outcomes. The Five Factor Score (FSS) and the Paediatric vasculitis activity score (PVAS) were calculated retrospectively to evaluate the disease activity. The results from the current study were compared with similar other studies across the world available in literature.

Study Procedures, data sources and measurements:

Participant records in the eMR were assessed for eligibility. The eMR personal helped retrieve the clinical data without any personal identifiers. The study team then extracted the relvant data using a data extraction form. The data were entered into Microsoft Excel (Publisher: Microsoft Corporation, Redmond, Washington, USA, 2016) on computers with password protection. Stringent measures were implemented to ensure the strict confidentiality of patient information throughout the process.

Bias:

A retrospective observational study is inherently susceptible to selection bias, especially referral bias [9, 10]. We posit that the referral bias resulting from the selective referral of a specific group of patients based on the variables of interest [10] was minimal, given that Bengaluru city, India, has twenty-one other medical colleges, comprising both public and private teaching institutes besides the innumerable corporate hospitals. Another potential concern in such studies is recall bias [9], but this is also expected to have been minimized as the data in the database were consistently recorded in real-time, and the current study was merely a pre-planned analysis carried out at a later date.

Patient and public involvement:

The development of the research question, study design, recruitment, reporting, and dissemination plans did not include input from patients or the public. While eligible individual eMR were included, these patients will not receive individual notifications of study findings. However, they have the option to request and access the results. Additionally, the outcomes will be disseminated through a scientific publication derived from the collected data.

Sample size estimation and statistical analyses:
There was no formal sample size estimation as the primary objective of the study was descriptive in nature and we proposed to include all eligible records available in the eMR between the years 2010 and 2022. The clinical features, laboratory, histopathology and radiological findings, pharmacotherapy and treatment outcomes were summarised using descriptive statistics. Statistical analyses were conducted utilizing Statistical Product and Service Solutions (SPSS) version 29 (IBM Corporation, Armonk, New York, 2022). Similar parameters from three other studies available in published literature were tabulated for comparison without any statistical analysis as the studies differed in their methodology and setting.

RESULTS:
Demography:
Our study included N=23 children diagnosed with cPAN with slightly male predominance (52% males). There were no exclusions. The mean (standard deviation [SD]) age of children affected was 8.78 (3.27) years. Four children were aged less than 5 years, while 13 of them were in the age group of 6 to 10 years and six children were aged more than 11 years.

Clinical presentation:
The most common clinical manifestations at diagnosis were fever (n = 22/23; 93%), skin manifestations, (n = 17/23; 74%) myalgia and arthralgia (n = 13/23; 56% each). The other clinical manifestations are summarized in Table1. The skin manifestations by decreasing order of frequency were purpura (n=18/22; 81.82%), subcutaneous nodule (n=10/22, 4.55%) livedo reticularis (n=7/22; 31.82%), skin ulcers (n=5/22; 22.73%), Raynaud's phenomenon (n=5/22; 22.73%), and skin gangrene (n=4/22; 18.18%). One of the patients with cPAN who had a mutation for DADA2 developed type 1 Diabetes mellitus subsequently. Short stature and hypothyroidism were noted in a couple of patients.

Table 1. Clinical Manifestations

Signs and symptoms	No. (%) of patients
Fever	22 (96%)
Skin manifestation	17 (74%)
Myalgia	13 (56%)
Arthralgia	13 (56%)
Weight loss	11 (48%)
Peripheral neuropathy	7 (30%)
Gastrointestinal involvement	7 (30%)
CNS involvement	4 (17%)
Renal involvement	4 (17%)
Testicular involvement	5 (22%)
Co morbidities	3 (13%)

Table 2. Laboratory Parameters

Laboratory parameter	n	Median (minimum, maximum values)	Mean ± Standard Deviation)
Haemoglobin	23	10.1 (2.5, 14.4)	10.71 (3.15)
Total count	23	17200 (37000, 5960)	16873.04 (8353.28)
Platelet count	23	420000 (840000, 220000)	465000 (90111)
C-Reactive Protein	18	13.4 (LLQ, 200)	Not calculable*
Erythrocyte Sedimentation Rate	20	70 (10, 138)	70.83 (31.86)

LLQ – Lower limit of quantification

*** Results have been issued as positive / negative for 8 patients with no values mentioned**

Table 3: Disease Activity Score

Particulars	Mean	SD	Median	Minimum value	Maximum value	Range

Five Factor score	0.69	0.80	1	0	3	3
Paediatric Vasculitis Activity Score	6.17	2.22	6	3	13	10

Table 4: Comparison of our study with other studies

Parameters	Current Study 2023 (N=23)	Lee et al, [6] 2021 (N=9)	Sönmez et al, [8] 2019 (N=133)*	Eleftheriou et al, [7] 2013 (N=69)
Country	India	South Korea	Turkey	United Kingdom
Median age of diagnosis in years Minimum – maximum age in years	8.78 (4-15)	7.6 (3-17.5)	NA (2-75)	8.5 (0.9-15.8)
Male: Female	1.1:1	2:1	1.4:1	1.22:1
Fever	22 (95%)	7 (77.7%)	86 (64.7%)	60 (87%)
Skin manifestations 1.Purpura Subcutaneous nodules Livedo reticularis Gangrene ulcers Raynauds phenomenon	18 (78%) 10 (43%) 7 (30%) 4 (17%) 5 (22%) 5 (22%)	8 (88.8%) 3 (33.3%) 2 (22.2%) 2 (22.2%) 2 (22.2%) 4 (44.4%)	NA NA 48 (36.1) 12 (9.0%) NA 13 (9.8%)	28 (41%) 16 (23%) 34 (49%) 3 (4%) 3 (4%) 16 (23%)
Musculo-skeletal manifestations Myalgia Arthralgia	13 (56%) 13 (56%)	2 (22.2%) 7 (77.7%)	79 (59.4%) 87 (17.3%)	57 (83%) 52 (75%)
Weight loss	11 (48%)	1 (11.1%)	56 (42.1)	44 (64%)
Gastrointestinal manifestation Abdominal pain Ischemia/ Perforation	5 (22%) 2 (9%)	2 (22.2%) 1 (11.1%)	32 (24.1%)	28 (41%) 3 (4%)
Renal manifestations Haematuria Impaired renal function	3 (13%) 2 (9%)	NA 1 (11.1%)	54 (40.6%)	7 (10%) 10 (15%)
Nervous system manifestation Stroke Peripheral neuropathy	3 (13%) 7 (30%)	1 (11.1%) 1 (11.1%)	43 (32.3%)	7 (10%) 6 (8.7%)
Testicular involvement	5 (22%)	NA	14 (10.5)	7 (10%)
Hypertension	3 (13%)	1 (11.1%)	54 (40.6%)	11 (16%)
Skin biopsy	15 (65%)	8 (88.8%)	94 (70.7%)	50 (72.4%)
Angiography	8(35%)	1 (11.1%)	92 (69.2%)	66 (95.6%)
Immunomodulators Corticosteroids Azathioprine	23 (100%) 16 (70%)	9 (100%) 4 (44.4%)	133 (100%) 31 (23.3%)	69 (100%) 54 (78%)

Cyclophosphamide Mycophenolate Mofetil	14 (60%) 8 (34%)	5 (55.5%) NA	62 (46.6) 39 (29.3)	60 (86.9%) 9 (13%)
Diagnosis Systemic PAN Cutaneous PAN Other vasculitides	 15 (65%) 8 (35%) NA	 8 (88.8%) 1 (11.1%) NA	 63 (47.4%) 33 (24.8%) 37 (27.8%)	 69 (100%) 0 (0%) NA
Mortality	2 (8.7%)	0	6(4.5%)	3 (4%)
Relapse	18 (78.2%)	7 (77.7%)	133 (100%)	24 (35%)
Median duration of follow up in years	5.4	7	13	6
FFS (Median)	1	2	NA	NA
PVAS (Median)	6	7	11*	NA

PAN – Polyarteritis Nodosa, FFS - Five Factor Score, PVAS - Paediatric vasculitis activity score

*** Includes n=67 adults except for PVAS which was solely from the paediatric population**

Laboratory parameters and Diagnosis:
All N=23 children had elevated acute phase reactants (ESR/CRP). Anemia, leukocytosis and thrombocytosis were seen in n=11/23 (47.83%); n=14/23 (60.87%) and n=8/23 (34.78%) respectively. The Anti-Streptolysin O (ASO) titres were elevated in n=6/23 (26.09%) of the patients. ANA and ANCA were tested is n=19/23 (82.61%) and n=16/23 (69.57%) patients respectively and none of them were positive for both. A summary of the laboratory parameters is given in Table 2. A total of n=15/23 (65.22%) patients were diagnosed via skin biopsy and the rest n=8/23 (34.78%) of them had abnormal angiography findings. With regards to DADA2 evaluation, all the five patients who were tested reported ADA2 gene mutation. Systemic PAN was diagnosed in n=15/23 (65.22%) children while the remaining n=8/23 (34.78%) had features of cutaneous PAN.

Pharmacotherapy and outcomes:
Prednsiolone was administered to all the N=23 children. Additional immunosuppression in the form of azathioprine, cyclophosphamide and mycophenolate Mofetil were given in n=16/23 (69.57%), n=14/23 (60.87%) and n=8/23 (34.78%) patients respectively. Benzathine penicillin prophylaxis was given in n=3/23 (13.04%) patients as they had elevated ASO titres.

With regards to the outcome, n=5/23 (21.74%) patients experienced no relapses during treatment, while n=11/23 (47.83%) patients encountered relapses while on treatment, and n=7/23 (30.43%) patients experienced relapses after the cessation of treatment. The FSS and PVAS are summarized in Table 3.

Comparison with other published studies:
Table – 4 summarises the demographics, clinical manifestations, pharmacotherapy and outcomes of the current study as well as three other studies. There was a slight male preponderance noted in all studies with Fever being the most common clinical manifestation followed by skin and musculoskeletal symptoms. Corticosteroids has been given in all patients from all the studies.

DISCUSSION:
We conducted a retrospective analysis of the eMR of patients with a diagnosis of cPAN who visited out institute over a period of 13 years (2010 – 2022). The clinical features, laboratory, histopathology and radiological findings, pharmacotherapy and treatment outcomes were summarised and then compared with other similar studies from South Korea, [6] Turkey [8] and United Kingdom [7]. Fever was the most common presenting complaint and all patients from all the 4 studies were administered corticosteroids for the induction phase in accordance with the current recommendations [3]. Relapse rate was 78.26 % (n=18/23) and mortality was seen in 8.7% (n=2/23).

All the four studies show a slight male preponderance as noted from previous epidemiological studies [11] and the median age of onset in the current study is approximately 9 years thereby corroborating the findings of Eleftheriou et al. [7] Although PAN is more common in the middle-aged and elderly age group it can affect any age and the median age of onset of cPAN is known to be approximately 10 years. [12] As per our routine practice, mutations for DADA2 were evaluated only in five children who presented with symptoms very early in life or refractory to treatment or presented with stroke and all of them were diagnosed to have DADA2 mutation.

The most prevalent clinical features of the disease at the time of diagnosis were fever, observed in 96% of patients, and skin involvement, noted in 74% of patients. Comparable results were reported in other studies as well [6 – 8]. Notably, Sönmez et al, have reported that skin manifestations are more common in children while symptoms of the nervous system are more common in the adults. [8] For instance, The French Vasculitis Study Group Database conducted a review of 348 adult patients with PAN (225 with PAN and 123 with Hepatitis B-PAN). Their findings also indicated that the most common observations in adults were constitutional symptoms (93.1%), with neurological findings following closely behind (79%). [13]

In accordance with the current guidelines issued in 2021 by the American College of Rheumatology (ACR) and the Vasculitis Foundation (VF) on the management of PAN, all patients were on Glucocorticoids and an additional immunosuppressive agent rather than glucocorticoids alone. In the case of cutaneous PAN patients, the standard treatment involved initiating therapy with oral or high-dose pulse intravenous corticosteroids, followed by maintenance treatment using low-dose prednisolone and a steroid-sparing agent, commonly azathioprine. For systemic PAN cases, the standard treatment regimen included induction therapy with high-dose pulse intravenous corticosteroids and intravenous pulse cyclophosphamide, followed by maintenance therapy with low-dose prednisolone and either azathioprine or Mycophenolate Mofetil. This treatment approach aligns with the EULAR recommendations for managing primary and medium-vessel vasculitis. [14]

With regards to the outcomes, the relapse rate was 78.26 % (n=18/23). The relapse rate seen in our study was similar to the one reported by Lee et al. [6] from South Korea (77.7%) and lesser than that reported by Sönmez et al. [8] from Turkey (100%). This could be attributed to the fact that Sönmez et al. had a long follow-up with a median of 13 years while the rest were for a much shorter duration of follow-up. Mortality in our study was 8.7% (n=2/23), which was higher than the reported in the other studies compared [6-8]. These variations could be attributed to the small sample size in our study as well as the other comparator studies. The mean FFS and PVAS score was 0.69 and 6.17, respectively which was similar to that reported by Lee et al. [6] Patients with higher PVAS had poor prognosis due to high disease activity.

The strength of our study is that it is one of the first studies with a notable number of patients with cPAN done from the Indian Subcontinent. However, our study has a few limitations. A retrospective study possesses inherent limitations, including biases stemming from underreporting, misclassification, and missing data. [15] To address these issues, conducting prospective studies in the future could help validate the findings of the current study. The sample size may still be considered small and a larger sample size including patients from multiple centres may be required for better generalizability. Moreover, databases typically lack routine in-depth collection of specific treatment details, challenges encountered during the medical management, and the severity of the complications, all of which can influence outcomes. Additionally, crucial information related to predictive factors such as the patient's functional status, affordability and access to health care were not available.

CONCLUSION:

cPAN is a rare disease of insidious onset and variable clinical presentation and is associated with severe complications despite appropriate treatment. Patients with cPAN most commonly present with constitutional symptoms followed by skin manifestations. Patients refractory to therapy, presentation at very young age, and presence of family history or stroke should be screened for ADA2 mutation. Although relapse is common, mortality is less than 10% if medical management was initiated in a timely manner. Thus, early diagnosis and aggressivetreatment for cPAN are important for improving prognosis and reduce the incidence of complications.

Declarations:

Ethics:

This study was approved by the Institutional Ethics Committee, Manipal Hospital vide letter A consent waiver was obtained as there was no patient interaction

Acknowledgements:

None

Conflicts of Interest:

None to Declare

REFERENCES:

1. Huang Z, Li T, Nigrovic PA, Lee PY. Polyarteritis nodosa and deficiency of adenosine deaminase 2 - Shared genealogy, generations apart. Clin Immunol. 2020 Jun;215:108411. doi: 10.1016/j.clim.2020.108411.
2. Iudici M, Quartier P, Pagnoux C, Merlin E, Agard C, Aouba A, Roblot P, Cohen P, Terrier B, Mouthon L, Guillevin L, Puéchal X; French Vasculitis Study Group. Childhood- versus Adult-Onset Polyarteritis Nodosa Results from the French Vasculitis Study Group Registry. Autoimmun Rev. 2018 Oct;17(10):984-989. doi: 10.1016/j.autrev.2018.08.001.

3. Ozen S, Ruperto N, Dillon MJ, Bagga A, Barron K, Davin JC, Kawasaki T, Lindsley C, Petty RE, Prieur AM, Ravelli A, Woo P. EULAR/PReS endorsed consensus criteria for the classification of childhood vasculitides. Ann Rheum Dis. 2006 Jul;65(7):936- 41. doi: 10.1136/ard.2005.046300.
4. Bae YD, Choi HJ, Lee JC, Park JJ, Lee YJ, Lee EB, Song YW. Clinical features of polyarteritis nodosa in Korea. J Korean Med Sci. 2006 Aug;21(4):591-5. doi: 10.3346/jkms.2006.21.4.591.
5. Navon Elkan P, Pierce SB, Segel R, Walsh T, Barash J, Padeh S, Zlotogorski A, Berkun Y, Press JJ, Mukamel M, Voth I, Hashkes PJ, Harel L, Hoffer V, Ling E, Yalcinkaya F, Kasapcopur O, Lee MK, Klevit RE, Renbaum P, Weinberg-Shukron A, Sener EF, Schormair B, Zeligson S, Marek-Yagel D, Strom TM, Shohat M, Singer A, Rubinow A, Pras E, Winkelmann J, Tekin M, Anikster Y, King MC, Levy-Lahad E. Mutant adenosine deaminase 2 in a polyarteritis nodosa vasculopathy. N Engl J Med. 2014 Mar 6;370(10):921-31. doi: 10.1056/NEJMoa1307362.
6. Lee JS, Kim JG, Lee S. Clinical presentations and long term prognosis of childhood onset polyarteritis nodosa in single centre of Korea. Sci Rep. 2021 Apr 16;11(1):8393. doi: 10.1038/s41598-021-87718-6.
7. Eleftheriou D, Dillon MJ, Tullus K, Marks SD, Pilkington CA, Roebuck DJ, Klein NJ, Brogan PA. Systemic polyarteritis nodosa in the young: a single-center experience over thirty-two years. Arthritis Rheum. 2013 Sep;65(9):2476-85. doi: 10.1002/art.38024.
8. Sönmez HE, Armağan B, Ayan G, Barut K, Batu ED, Erden A, Uğurlu S, Bilginer Y, Kasapçopur Ö, Karadağ Ö, Bilgen SA, Özdoğan H, Ozen S. Polyarteritis nodosa: lessons from 25 years of experience. Clin Exp Rheumatol. 2019 Mar-Apr;37 Suppl 117(2):52-56.
9. Jager KJ, Tripepi G, Chesnaye NC, Dekker FW, Zoccali C, Stel VS. Where to look for the most frequent biases? Nephrology (Carlton). 2020;25(6):435-441.
10. Sutton-Tyrrell K. Assessing bias in case-control studies. Proper selection of cases and controls. Stroke. 1991 Jul;22(7):938-42. doi: 10.1161/01.str.22.7.938. PMID: 1853415.
11. Mahr A, Guillevin L, Poissonnet M, Aymé S. Prevalences of polyarteritis nodosa, microscopic polyangiitis, Wegener's granulomatosis, and Churg-Strauss syndrome in a French urban multiethnic population in 2000: a capture-recapture estimate. Arthritis Rheum. 2004 Feb 15;51(1):92-9.
12. Bansal NK, Houghton KM. Cutaneous polyarteritis nodosa in childhood: a case report and review of the literature. Arthritis. 2010;2010:687547. doi: 10.1155/2010/687547.
13. Pagnoux C, Seror R, Henegar C, Mahr A, Cohen P, Le Guern V, Bienvenu B, Mouthon L, Guillevin L; French Vasculitis Study Group. Clinical features and outcomes in 348 patients with polyarteritis nodosa: a systematic retrospective study of patients diagnosed between 1963 and 2005 and entered into the French Vasculitis Study Group Database. Arthritis Rheum. 2010 Feb;62(2):616-26. doi: 10.1002/art.27240.
14. Mukhtyar C, Guillevin L, Cid MC, Dasgupta B, de Groot K, Gross W, Hauser T, Hellmich B, Jayne D, Kallenberg CG, Merkel PA, Raspe H, Salvarani C, Scott DG, Stegeman C, Watts R, Westman K, Witter J, Yazici H, Luqmani R; European Vasculitis Study Group. EULAR recommendations for the management of primary small and medium vessel vasculitis. Ann Rheum Dis. 2009 Mar;68(3):310-7. doi: 10.1136/ard.2008.088096.
15. Mann CJ. Observational research methods. Research design II: cohort, cross sectional, and case-control studies. Emerg Med J. 2003;20:54–60.