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Case Report

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Cervical Syringomyelia in a Bechet Disease with SLE-associated Clinical Symptoms: A Case report

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Abstract

Behçet's disease, an inflammatory disorder with oral and genital ulcers and ocular involvement, rarely presents with syringomyelia, characterized by cystic spinal cord expansion. A 39-year-old female with Behçet's disease developed severe neck pain radiating into her arms. Despite ongoing treatment with hydroxychloroquine, colchicine, and pain management, MRI revealed cervical syringomyelia. Conservative management was insufficient, necessitating referral to a tertiary neurosurgeon. This case illustrates the diagnostic and management challenges of overlapping autoimmune and neurological conditions. It emphasizes the need for a multidisciplinary approach and ongoing treatment adjustments. Managing Behçet's disease complicated by syringomyelia requires an integrated strategy and careful monitoring to optimize patient outcomes.

Keywords: Vasculitis, Autoimmune, Bechet's Disease, Systemic lupus erythematosus, Syringomyelia

Introduction

Autoimmune condition happens when protective mechanism accidently starts targeting its own body parts including vessels. Involvement of autoimmune mechanism to blood vessel leads to inflammation of blood vessels which knows as vasculitis. Vasculitis involves range of blood vessels from small to large vessels of body [1], which presents with vast clinical symptoms and categories as different form of vasculitis based on vessel involved [2] [3]. The annual incidence of vasculitis as per studies is approximately 38 to 40 people per million and is more common in adults compare to the children [4]. There are lists of vasculitis and one of them is Bechet's disease. Bechet's disease is a chronic multisystemic inflammatory disorder also known as the Silk Route disease, which presents with numerus symptoms mainly with a triad of symptoms includes oral aphthous ulcers, genital ulcers, and ocular disease [5][6][13]. Excluding other autoimmune as SLE is important through a workup as symptoms of Bechet's disease can be overlap with the symptoms of other autoimmune disorders with spectrum of clinical presentations. Bechet's disease is a diagnosis of exclusion [7]. HLA-B5 and HLA-B51 are gene markers that are sometimes present in patients with Behçet's disease [8]. However clinical presentation varies from individual to another and can present as a rare symptom, which requires documentation and presentation.

Case Presentation

A 39-year-old married female, a non-smoker and non-drinker, allergic to Amoxicillin Trihydrate and potassium clavulanate (Augmentin), was referred to the Arthritis Association of Kingsport, PLLC by an NP rheumatologist on

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04/06/2020 at 10:00 a.m. The referral was due to abnormal lab results and arthralgia. The patient's primary physician was concerned about lupus, neuropathy, and a borderline positive ANA (1:160). Two years prior, the patient had reported weight gain and frequent throat clearing. An evaluation revealed a spot on her thyroid, which was later diagnosed as a cyst. She subsequently experienced GI upset, nausea, and vomiting, and was suspected to have pelvic congestion syndrome, a diagnosis later doubted by her gynecologist. An endoscopy was performed, which was negative for celiac disease and colitis. An MRI of the head was done which was normal. The NP rheumatologist was changed, and the patient was prescribed Plaquenil (hydroxychloroquine) 200 mg BID. At the time of presentation to the Arthritis Association of Kingsport, PLLC, the patient stated, “I feel like I am falling apart.” She reported severe fatigue, twitching, spasms, and a tendency to develop a severe rash with white spots on her face when exposed to the sun. She has also experienced clear fluid-filled blisters on her scalp and stated that her teeth have been breaking apart. Her mouth felt dry, her eyes were itchy, and she experienced diplopia. Additionally, she had occasional oral and nasal ulcers, dizziness upon standing, and some morning stiffness. Her medications included norethindrone (progestin) once daily, gabapentin once daily, dicyclomine once daily, and Elavil (amitriptyline) at bedtime, in addition to Plaquenil. Her initial vitals were recorded at 10:00 a.m. (see Tablet *1*). A detailed systemic review was performed, which was positive for mouth ulcers, nasal sores, dyspnea, chest pain, extremity weakness, gait disturbance, numbness in extremities, tingling, and skin lesions. Physical examination revealed several small raised maculopapular lesions in malar regions, upper limb digits had hypermobility, painful shoulders at full rom, hyper clonus in the feet, DTRs symmetrically increased and had mild soreness in L- spine region.

Table 1: vital signs recorded on 4/6/2020

MEASUREMENT	PATIENT'S VALUE	NORMAL RANGE
Blood pressure (mm Hg)	120/80 mmHg	120/80
Pulse (pulse per min)	84/min	60-100
Respiratory rate (breathes per min)	18/min	Dec-20
Temperature (F)	97.9 F	97-99
Weight (lb)	162	Variable
Weight (kilogram)	73.4	Variable
Height (foot)	5	Variable
Height (centimeter)	165	Variable
Body Mass Index (kg/m2)	26.96	18.5-24.9

mmHg (millimeters of mercury): A unit of pressure often used to measure blood pressure and atmospheric pressure.

lb (pound): A unit of weight in the imperial system, commonly used to quantify mass in the United States.

F (Fahrenheit): A temperature scale where water freezes at 32°F and boils at 212°F, primarily used in the U.S.

kg/m² (kilograms per meter squared): A unit of measurement for density or body mass index (BMI), indicating mass per unit area.

Urinalysis was performed and found to be normal (see Table *2*). Additionally, the following tests were conducted: C-reactive protein (CRP), CBC, comprehensive metabolic panel (CMP), creatine kinase, erythrocyte sedimentation rate (ESR), rheumatoid factor (RF), ANA, uric acid, complete auto-antibody panel (ANA, IgG by IFA, Complement C3,Complement C4, dsDNA Antibodies, F-actin smooth muscle antibody, IgG, M2 anti-bodies, IgG by ELISA, Ribosomal P Antibodies, SS-A, SS-B,SCL-70,Sm/RNP Antibodies) and thyroid studies (see Table *3*). She was started on a trial of colchicine 0.6 mg once a day, following detailed counseling including neurological assessment at some point and was advised on pain management for pain intensity rated at 3/10. A follow-up was planned for one month later. On 02/08/2021 at 08:15 a.m., the patient returned for her Behçet's disease follow-up (Onset date: 24/09/2020) and medication monitoring. She reported that she was doing well on Mitigare (colchicine) 0.6 mg and only had occasional oral ulcers. Additionally, she experienced migraines twice a week, which improved with dietary changes. She also had blisters behind her left ear and along the lower jawline, which were likely due to the elastic in her mask; these improved as she reduced her mask use. She had an ear infection a few weeks prior to the follow-up, which was cleared with antibiotics. Her previous studies were reviewed, and CBC, ESR, CRP, and biochemistry results were normal. Functional limitations were assessed: she was able to get in and out of a car, go down stairs, perform activities of daily living, put on socks and shoes, and walk. However, she found it difficult to climb stairs, exercise, kneel, and walk more than 5-10 blocks or cover unlimited distances. In addition to her previous medications, she was taking dicyclomine 10 mg twice a day, gabapentin 300 mg once daily, and norethindrone acetate 5 mg once daily during the second half of her menstrual cycle. Vitals were recorded at 8:23 am (see Table *4*). On physical examination, she had scarring from blisters behind her left ear and some red lesions along the lower mandible. She was maintained on the same medications, and pain management was advised. A fall risk assessment was performed, which indicated no risk of falling. She later developed significant neck problems and presented on 11/4/2022 at 8:00 a.m. She reported neck pain radiating into both arms. she received multiple steroid injections for neck pain. Two MRIs of the cervical spine were performed, revealing a syrinx.

She consulted with a neurosurgeon, who increased her pain medications and opted for conservative management without any surgical intervention. The impression was made, that the cervical syringomyelia was not associated with

Table 2: Urinalysis

COMPONENT	RESULTS	NORMAL RANGES
Glucose	Negative	≤130 mg/d
Blood	Negative	≤3 red blood cells
Protein	Negative	≤150 mg/d
Nitrate	Negative	Negative
Leukocyte	Negative	≤2–5 WBCs/hpf

Table 3: LABs collected on 04/06/2020

LABS	RESULTS	NORMAL VALUES	UNIT
Hemoglobin	9.3	11.5-15.5	Gm/dL
Hematocrit	30.9	35.2-46.4	%
Platelet counts	324	137-397	K/cumm
Total leukocytic count	7.98	3.8-11.5	K/uL
Erythrocyte Sedimentation Rate (ESR)	45	<26	mm/hr
Albumin/Globulin ratio	2	1.1-2.5	Mg/dl
Albumin	4.5	3.5-5.2	g/dl
Alkaline phosphatase	101	35-121	IU/L
Serum glutamic pyruvic transaminase	32	<5-40	IU/L
Bilirubin, Total	0.7	<0.2-1.2	mg/dL
blood urea nitrogen (BUN)	19	Jun-20	Mg/dl
Calcium	9.9	8.6-10.4	Mg/dl
Chloride	100	97-108	Mmol/L
Creatinine	0.9	0.5-1	Mg/dl
Potassium	4.3	3.5-5.3	Mmol/L
Sodium	138	135-145	Mmol/L
Creatine kinase	14	20-180	U/L
C-Reactive protein	7	<0.5	Mg/dl
Vitamin-D (25-hydroxy)	46	30-100	Ng/dl
Thyroid stimulating hormone (TSH)	2.3	0.43-5.2	mU/L
Free thyroxine 4 (Ft4)	1.1	0.8-1.7	ng/dL
Complement C3	190	90-180	mg/dL
Complement C4	29	Oct-40	mg/dL
Hepatitis B core antibodies (HBcAb)	Non-reactive	Non-reactive	
Hepatitis B surface antibodies (HBsAb)	Non-reactive	<10 = non-reactive	
Hepatitis B surface antigen (HBsAg)	Non-reactive	Non-reactive	
Hepatitis C antibodies IgG	Non-reactive	Non-reactive	
RA factor	10	< 14	IU/mL
ANA	0.152777778	< 1:40	Dilution
IgG by IFA	657	700-1600	Mg/dl
DsDNA ANTIBODY	20	< 30	IU/mL
F-actin smooth muscle antibody, IgG	01:10	< 1:20	dilution

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M2 anti-bodies, IgG	0.166666667	< 1:20	dilution
Ribosomal P Antibodies	01:10	< 1:20	dilution
SS-A	Negative	Negative	
SS-B	Negative	Negative	
SCL-70	Negative	Negative	
Sm /RNP Antibodies	Negative	Negative	
Uric acid	3.5	2.4 to 6.0	mg/dL

mg/dL (milligrams per deciliter): Measures concentration in blood, commonly for glucose and cholesterol.
IU/mL (international units per milliliter): Standardized measure for biological activity of substances.
AU/mL (arbitrary units per milliliter): Relative unit based on reference standards in diagnostics.
mmol/L (millimoles per liter): Indicates concentration of solutes, used for blood metabolites.

Table 4: vital signs recorded on 2/8/2021

MEASUREMENT	PATIENT'S VALUE	NORMAL RANGE
Blood pressure (mm Hg)	110/82 mmHg	120/80
Pulse (pulse per min)	83/min	60-100
Respiratory rate (breathes per min)	16/min	Dec-20
Temperature (F)	97.7 F	97-99
Weight (lb)	162	Variable
Weight (kilogram)	73.4	Variable
Height (foot)	5	Variable
Height (centimeter)	165	Variable
Body Mass Index (kg/m2)	26.96	18.5-24.9
Oxygen saturation	98%	>92%

mmHg (millimeters of mercury): A unit of pressure often used to measure blood pressure and atmospheric pressure.
lb (pound): A unit of weight in the imperial system, commonly used to quantify mass in the United States.
F (Fahrenheit): A temperature scale where water freezes at 32°F and boils at 212°F, primarily used in the U.S.
kg/m² (kilograms per meter squared): A unit of measurement for density or body mass index (BMI), indicating mass per unit area.

Table 5: List Of Medications

MEDICATION	DOSE	FORM	FREQUENCY
Dicyclomine	10 milligrams	Capsule	Twice per day
Gabapentin	300 milligrams	Capsule	Once per day
Plaquenil	200 milligrams	Capsule	Twice per day
Colchicine	0.6 milligrams	Tablet	Once per day
Amitriptyline	10 milligrams	Tablet	Once at bedtime
Norethindrone acetate	5 milligrams	Tablet	Once per day At 2nd half of menstrual cycle
Mitigare	0.6 milligram	Capsule	Twice per day

Behçet's disease or not aware of the link. It was suggested that a neurosurgeon from a tertiary care hospital, who had experience performing decompression in such cases, be consulted as previous neurosurgeon is unaware of this new diagnosis. She had done well from Bechet's standpoint. She still had some lesion on her face related to the mask, which flare up along with generalize disease flare. Functional limitations were same as before. She was being managed on syringomyelia (onset date: 11/4/2022) and Bechet's disease (onset date: 24/09/2020). medications were kept as before (table 5).

Vitals were recorded (see table 6). systemic review and physical examination were normal. Pain score of 6/10. The neck pain has adversely affected her ability to function and exercise. Bechet's was stable. Medications were continued and pain management was advised. Her neck pain was developed while she has been on colchicine. Neurologist visit was advised for assessment and alternative medication to suggest. The exact cause of syringomyelia is not clear.

Table 6: vital signs recorded on 11/2/2022

MEASUREMENT	PATIENT'S VALUE	NORMAL RANGE
Blood pressure (mm Hg)	130/82mmHg	120/80
Pulse (pulse per min)	87/min	60-100
Respiratory rate (breathes per min)	16/min	Dec-20
Temperature (F)	97.7 F	97-99
Weight (lb)	165	Variable
Weight (kilogram)	79.8	Variable
Height (foot)	5	Variable
Height (centimeter)	165	Variable
Body Mass Index (kg/m2)	29.2	18.5-24.9
Oxygen saturation	98%	>92%

mmHg (millimeters of mercury): A unit of pressure often used to measure blood pressure and atmospheric pressure.

lb (pound): A unit of weight in the imperial system, commonly used to quantify mass in the United States.

F (Fahrenheit): A temperature scale where water freezes at 32°F and boils at 212°F, primarily used in the U.S.

kg/m² (kilograms per meter squared): A unit of measurement for density or body mass index (BMI), indicating mass per unit area.

Discussion

We presented the case report, which presents a complex interplay of Behçet's disease, syringomyelia, and various symptoms in a 39-year- old female patient. The multifaceted nature of her symptoms and treatment highlights several important aspects of managing chronic systemic diseases and their complications. The patient's presentation with Behçet's disease, characterized by recurrent oral and nasal ulcers, rash, and neurological symptoms, alongside the recent diagnosis of syringomyelia, underscores the challenges in diagnosing and managing patients with overlapping or multifactorial conditions. Behçet's disease can present with a wide range of symptoms [9], including skin lesions, oral ulcers, and systemic manifestations, which can complicate the diagnostic process. The addition of syringomyelia, a condition that involves cystic enlargement within the spinal cord, introduces further complexity, as its symptoms and management can be distinct from those of Behçet's disease. The patient's medication regimen includes Plaquenil (hydroxychloroquine), colchicine, gabapentin, amitriptyline, dicyclomine, and norethindrone acetate. Each medication serves a specific purpose, Plaquenil is effective in controlling inflammatory symptoms of Behçet's disease, Colchicine targets oral ulcers and systemic inflammation associated with Behçet's, Gabapentin and amitriptyline are employed for neuropathic pain management, which is crucial given the patient's reports of severe fatigue, pain, and neurological symptoms, Dicyclomine addresses gastrointestinal symptoms, while norethindrone acetate helps manage menstrual cycle-related symptoms [10] The presence of multiple medications necessitates careful monitoring for drug interactions and side effects. For instance, the potential overlap in effects between colchicine and Mitigare (another formulation of colchicine) needs to be scrutinized. Additionally, the impact of gabapentin and amitriptyline on cognitive function and pain management should be evaluated [11], especially in the context of a patient with significant neurological symptoms. The development of syringomyelia, confirmed by MRI, has significantly impacted the patient's functional capacity [12]. Symptoms such as neck pain radiating into the arms, along with the diagnosis of syringomyelia, necessitate an evaluation of its potential impact on the patient's quality of life and functional abilities. Despite conservative management by the neurosurgeon, the significant pain and functional limitations experienced by the patient underscore the need for ongoing assessment and possibly advanced surgical options. The decision for conservative management rather than immediate surgery may be based on the current understanding of the syringomyelia's progression and its association with Behçet's disease.This case emphasizes the importance of an interdisciplinary approach to patient care. The involvement of rheumatologists, neurologists, and other specialists is crucial for managing the overlapping symptoms of Behçet's disease and syringomyelia. Coordinated care ensures that all aspects of the patient's condition are addressed, from the management of chronic disease symptoms to the treatment of specific complications such as syringomyelia.The patient's evolving symptoms and treatment responses highlight the need for a personalized and dynamic approach to management. Regular follow-up and reassessment of the treatment plan are essential to address new symptoms or complications as they arise. This case underscores the importance of individualized care plans

that adapt to the patient's changing condition and treatment responses and highlights the need for further research into the relationship between Behçet's disease and syringomyelia. Understanding whether and how these conditions may be related could inform future management strategies. Additionally, exploring advanced treatment options, including surgical interventions and novel pharmacological therapies, may provide new avenues for improving patient outcomes.

Conclusion

This case report illustrates the complex management of a patient with Behçet's disease complicated by the development of syringomyelia. The patient's journey highlights the challenges of diagnosing and treating multifactorial conditions with overlapping symptoms. Despite the treatment regimen, which included medications such as Plaquenil, colchicine, gabapentin, and amitriptyline, the emergence of syringomyelia introduced significant new challenges. The patient's persistent neck pain and functional limitations despite conservative management underscore the need for an interdisciplinary approach to care. The case underscores the importance of a personalized, dynamic treatment strategy that evolves with the patient's symptoms and responses to therapy. Ongoing evaluation and adjustment of the treatment plan are crucial for addressing new symptoms and optimizing patient outcomes. Additionally, this case points to the need for further research into the potential links between Behçet's disease and syringomyelia, which could provide insights into more effective management strategies and contribute to a better understanding of these complex conditions. By coordinating care among specialists and closely monitoring the patient's condition, healthcare providers can better navigate the intricacies of managing such multifaceted cases, ultimately aiming to enhance the patient's quality of life and functional capabilities.

Patient Consent

A written informed consent was obtained from the patient for publication of this case report and any accompanying images.

Conflict of Interest

The authors declare that they have no conflicts of interest.

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