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Case Report

"Ten" Differentials for this "Gen" like Mass at Nape of Neck

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Abstract

Neck swellings/masses are one of the most common encounters in the ENT outpatient department. As a known fact, fascial planes in the neck are dense and deep containing rich network of arterial, venous and nerve supply which bridges head with the rest of the body. Hence, it is important to assess any simple to complicated neck swellings which have a diverse origin as well as varied differential. It is crucial to remember that early and timely intervention of any neck swellings will lead to apt and prompt management. Here, is one such "GEN" like swelling over the nape of the neck which is (Gigantic, Erratic, Necrotic) encountered at the ENT outpatient department which was definitely one of a kind

Keywords: Neck swellings; necrotic; hyperpigmentation; differential

Introduction

Neck swellings are encountered at all ages with an extensive and exclusive differential diagnosis ranging from a congenital mass to a cancerous growth. It is hence significant that we surgeons have an in-depth knowledge about the root of its origin that is the embryology and anatomy of the swelling so as to aid a pertinent conclusion and provide applicable management algorithm.

Case Report

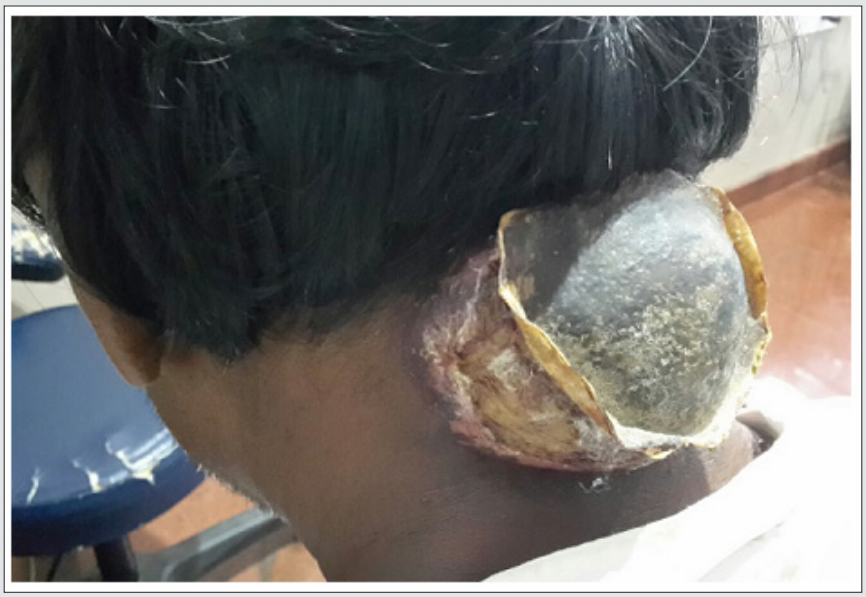

Figure 1:

(Figure 1) Here, is this patient who consults the ENT outpatient department on a regular basis with this huge swelling over the nape of the neck since 6 months. The growth was initially small in size (smaller than a lemon, now to have attained more than the size of the orange roughly) about 2 years back but has begun increasing in its size over a period of 6 months now. He also gives history of foul-smelling discharge along with peeling of skin from over the growth. There is pain on head and neck movements, heaviness around the neck, disturbances while sleeping and difficulty in taking shower. Patient has consulted at various places and tried his hands at various modes of treatment, with no symptomatic relief and barely satisfactory results. Patient ended up at this Tertiary care hospital on somebody's suggestion regarding good facilities and treatment care available at this place. He also gives history of compromised social and family life since the time of origin of this swelling; making him feel lonely, depressed and abandoned. A farmer by occupation who is with bare minimum financial build-up has now got habituated to smoking and consuming alcohol. Patient is a known case of both Hypertension and Diabetes (due to high levels of stress) and is on treatment for the same, which is said to be under control. His relatives have also mentioned on him taking antidepressants and sedatives due to the depression he is with off-lately. They have also mentioned hospital admissions several times due to alcohol withdrawal symptoms (as he is having chronic alcohol dependency). They have also hinted that he is having suicidal tendencies and tried to commit suicide at few times.

On general physical examination, he is normal built and nourished and oriented to time, place and person but showing signs of being depressed in life and living. His vitals were stable. There are hesitant marks on both of his forearm near the wrist. Systemic examination was within normal limits. There were no positive clinical findings on local examination of Ear, Nose and Throat. On local examination of neck- 5X5 cm huge, irregular, necrotic, firm to hard, foul-smelling mass over the nape of the neck, which was non-tender, adherent to the base while surface showing hyperpigmented, peeling skin with irregular borders and margins. The swelling was immobile in both horizontal and vertical directions and there was no bleed on touch. There was no induration around the swelling but with certain area of hyperpigmentation all around it. There was bleeding seen when tried to peel off the superficial hyperpigmented skin from over the swelling. There were no pulsations over and around the swelling plus there was no engorgement of veins noted over the swelling. Biopsy was planned under local examination. But, when explained to the patient the possible next line of treatment is based on the histo-pathological report, the patient was not willing for the same as he wanted relief only through symptomatic line of treatment. Following this OPD visit, the patient did not follow up. But, looking at this swelling there are numerous differentials that cross the mind which possibly could be the final diagnosis in this case which have been discussed below.

Discussion

There are various swellings occupying the head and neck region ranging from infective to neoplastic. As per this case report, the top 10 most likely swellings at the nape of neck that can be taken as probable differentials are elaborated below: Necrotising fasciitis (NF) secondary varicella zoster infection, Acne Keloidalis nuchae(AKN), Pyoderma gangrenosum(PG), Erysipelas, Gas gangrene(GG), Calciphylaxis, Ecthyma gangrenosum(EG), Purpura fulminans complicating varicella, Vibrio vulnificus(VV) infection, Subcutaneous acute febrile neutrophilic dermatosis (subcutaneous Sweet syndrome). Necrotizing fasciitis [1,2] is an infection that spreads along fascial planes causing subcutaneous tissue death characterized by rapid progression, systemic toxicity and even death. It appears as red, hot, painful and swollen wound with an ill-defined border. As the infective process continues, local pain is replaced by numbness or analgesia. As the disease process continues, skin initially becomes pale, mottled & purple and finally gangrenous. It then moves rapidly along fascial planes and causes tissue necrosis; secondary to its polymicrobial composition and synergistic effect of enzymes produced by bacteria. Treatment involves securing airway, broad-spectrum antimicrobials, intensive care support and prompt surgical debridement. Reducing mortality rests on early diagnosis and prompt aggressive treatment.

Acne Keloidalis nuchae [3-7] is a chronic inflammatory condition that leads to scarring of hair follicles, development of keloid-like papules and plaques, scarring alopecia on nape of the neck and occipital scalp. The incidence is between 0.45- 9%, occurring mostly in darker skinned races with curly or kinky hair. It is most common in blacks and in Caucasians with M: F = 20:1 and starts after adolescence. The natural course of disease starts with early formation of inflamed papules with marked erythema [3]. Secondary infection can lead to pustules and abscess formation. This continued inflammation leads to pronounced fibrosis and keloid formation with coalescence of papules into plaques and later nodules. The exact underlying pathogenesis is not known but two predominant theories suggest skin injury and existence of aberrant immune reactions. Skin injuries from irritation, occlusion, trauma, friction and hair cutting practices are all risk factors. It has been proposed that due to an immune reaction that can lead to cicatrical alopecia [5,7]. On histology, there is mixed, neutrophilic, lymphocytic infiltration. In this theory, intrafollicular antigens attract inflammatory cells to the follicle resulting in damage to sebaceous gland and follicular wall. This in turn leads to rupture of follicle and release of antigens into hair follicle that precipitates inflammatory process and epithelial destruction leading to fibrosis [4]. Traditional management focuses on preventing disease progression, avoidance of mechanical irritation from clothing and use of antimicrobial cleansers to prevent secondary infection. Treatment involves use of topical, intralesional or systemic steroids in combination with retinoids and/or antibiotics to decrease inflammation [7]. When disease progresses from early to late stage, surgical excision and skin grafting may be performed which require long periods of healing. Recent advances in light and laser therapies [3] offer an alternative treatment option. This condition matches the features of the patient in this study. Pyoderma gangrenosum [8,9] is a serious, progressive, necrotising, ulcerative skin disorder usually associated with underlying systemic diseases such as

ulcerative colitis, arthritis, polycythemia vera. Head and neck involvement is rare, but possibly more common. The etiology is unclear but may be due to abnormal immunologic response [8]. There are no pathognomonic histologic or laboratory findings; the diagnosis is made by the clinical appearance of lesions and disease course. Many management regimens have been recommended, but treatment must be individualized and must include concern for the physical and psychologic needs of the patient [9]. This particularly stressful and uncertain lesion is a challenge to both surgeons and physicians. Treatment is usually non-surgical and consists of immunosuppression and local wound care of ulcers in addition to find underlying primary systemic disorder. Systemic and intralesional steroids are the treatment of choice.

Erysipelas [10,11] is an infection of superficial layers of skin. The most common cause is group A streptococcal bacteria, especially Streptococcus pyogenes. Erysipelas results in a fiery red rash with raised edges that can easily be distinguished from the skin around it. The affected skin may be warm to touch. It was thought to affect mostly face, but off-lately the distribution of inflammation is changing where in legs are involved in 80% of cases. The lesion is seen affecting adults between 60-80 years of age also seen occurring in infants as well. The rash may also appear on the arms or trunk. Erysipelas [10,11] begins at the site of minor surgery or trauma: bruise, burn, wound, or incision or it may be due to lymphatic obstruction. When rash appears on the trunk, arms, or legs, it is usually at the site of a surgical incision or a wound. It first appears as a localized lesion that is tender and red with accompanying high fever, chills, headache, nausea and malaise. The skin in the affected area resembles peel of an orange [10]. The lesion quickly develops a bright red, shiny color with a spreading, raised border. The typical lesion is so characteristic that its presence is diagnostic. The treatment of choice is penicillin. For the penicillin-allergic patient, erythromycin or cephalexin may be used [11]. Gas gangrene [12,13] is a very rare but life-threatening necrotizing soft tissue infection (NSTI) caused by anaerobic, spore-forming, and gas-producing clostridium subspecies. It is the most rapidly spreading and lethal infection in humans, also affecting muscle tissue. The high mortality mediated by potent bacterial exotoxins. Gas gangrene common in the extremities is a relatively rare affliction of the head and neck [12]. The involvement of neck spaces as a sequel to acute pharyngolaryngeal inflammation, dental causes, trauma and unknown etiology. The causative organism is Clostridium perfringens in 80–95% of cases. Gas gangrene can rapidly progress to serious morbidity and even mortality, and prompt surgical intervention is clearly indicated [13]. Management of non-clostridial gas gangrene [12,13] of the neck is a challenge for the surgeons which houses major vessels and vital structures and in ensuring complete drainage and at times surgical debridement.

But recently recommended use of hyperbaric oxygen therapy in tandem with antibiotics for soft tissue infections. Gas gangrene [12,13] of the neck needs to be diagnosed quickly and managed aggressively for desirable results to follow. However, they warrant prompt recognition and immediate treatment because of their potential risk of rapid spread to the deep neck spaces, thrombophlebitis, mediastinitis and cardiac tampanode. Calciphylaxis [14,15] is a serious, uncommon disease in which calcium accumulates in small blood vessels of the fat and skin tissues. It causes blood clots, painful skin ulcers and may cause serious infections that can lead to death. The exact cause of calciphylaxis is unknown, but recent studies have revealed that most people with the condition have abnormalities in blood-clotting factors. People with calciphylaxis have an imbalance in the metabolism of calcium [14]. This causes calcium to be deposited in the smallest parts of the arteries (arterioles), which eventually leads to the formation of blood clots in the arterioles. Blood clots can cause fat tissues and skin to be deprived of oxygen and nourishment. Calciphylaxis most commonly affects people who have end-stage kidney failure [15]. Possible risk factors include female, Obesity, Diabetes, Abnormalities in blood-clotting factors, Long-term dialysis and sometimes kidney transplantation, have kidney failure and are on dialysis or have had a kidney transplant, imbalance of calcium, phosphorus and aluminum in the body, medications (warfarin, calcium-binding agents or corticosteroids), Hyperparathyroidism, Uremia. Surgical approach for calciphylaxis [14,15] can stimulate the sympathetic nerves and may cause the formation of new ulcers. Therefore, surgery may worsen the calciphylaxis prognosis, suggesting that surgical treatment should be selected with caution after careful consideration of the risks to the patient's condition and general risks. Currently, treatment for calciphylaxis [14,15] involves whole-body management, including prevention of infection, correction of the abnormal metabolism of calcium and phosphate, local management of ulcers by debridement. In recent years, treatment with sodium thiosulfate has been gaining attention. Cinacalcet acts directly on the calcium receptors in the parathyroid cells and inhibits parathyroid hormone without elevating blood serum calcium levels. It also lowers blood serum phosphate levels.

Ecthyma gangrenosum [16,17] is a rare, invasive, cutaneous lesion frequently associated with Pseudomonas aeruginosa, although it may develop in the absence of bacteremia and may originate from other bacterial and fungal organisms. It is most often occurring in patients with neutropenia and immunocompromised hosts. It typically occurs on the extremities, gluteal and perineal regions. It can also occur in Head and Neck region also [16]. Tissue and blood cultures will be positive for P aeruginosa which are diagnostic. This clinical entity should be considered when otolaryngologists are asked to evaluate necrotic cutaneous lesions of the head and neck. Treatment is usually broad spectrum antibiotics-Amikacin. Considering high rate of mortality, early diagnosis and prompt effective treatment is mandatory [17]. Purpura fulminans [18,19] is a rare complication after a primary infection with varicella zoster virus. A varicella infection may lead to protein C or S deficiency resulting in diffuse intravascular coagulation and severe skin defects. They usually occur after about 2 weeks after chickenpox infection. They present with large painful ecchymotic, necrotizing and retiform plaques on the lower extremities and also

in the Head and neck region. Because of the rapidly progressing purpura, patient presents with clinical signs of hypovolaemic shock [18]. Laboratory analyses revealed very low protein S levels as well as anticardiolipin antibodies. Aggressive treatment by low-molecular-weight heparin, steroids, intravenous immunoglobulins, fresh frozen plasma, prednisolone, acyclovir and broad-spectrum antibiotics- ceftriaxone was able to prevent the extension of the lesions and to correct the coagulation abnormalities [19].

Vibrio vulnificus [20,21] infection is a rare and causes severe illness including necrotising soft tissue infection, septicaemia, gastroenteritis having high mortality. The risk factors include immunocompromised state, diabetes and cirrhosis. Infection may be contracted by consuming undercooked seafood or swimming in contaminated sea water. V. vulnificus [20,21] is usually susceptible to tetracyclines alone, but adding a cephalosporin is associated with better therapeutic response. Tetracycline with or without cephalosporin is usually sufficient to treat this potentially lethal Gram-negative bacterium. Sweet's syndrome [22] is uncommon skin condition characterized by fever, neutrophilia, tender erythematous, painful skin lesions that appears on the arms, face, neck and diffuse infiltrate consisting mature neutrophils located in the upper dermis. The cause of Sweet's syndrome isn't known, but it's sometimes triggered by an infection, illness or medication. Sweet's syndrome presents in three clinical settings: classical (or idiopathic), malignancy-associated, and drug-induced. Classical Sweet's syndrome (CSS) [22] presents in women between 30 to 50 years of age, it is often preceded by an upper respiratory tract infection and may be associated with inflammatory bowel disease and pregnancy. 1/3rd with CSS experience recurrence of dermatosis. Malignancy-associated Sweet's syndrome (MASS) [22] can occur as a paraneoplastic syndrome in patients with established cancer or individuals whose Sweet's syndrome-related hematologic dyscrasia or solid tumor was previously undiscovered; MASS is most commonly related to acute myelogenous leukemia.

The dermatosis can precede, follow, or appear concurrent with the diagnosis of the patient's cancer. Hence, MASS can be the cutaneous harbinger of either an undiagnosed visceral malignancy in a previously cancer-free individual or an unsuspected cancer recurrence in an oncology patient. Drug-induced Sweet's syndrome (DISS) [22] occurs in patients who have been treated with granulocyte-colony stimulating factor. The pathogenesis of Sweet's syndrome may be multifactorial and still remains definitively established. Clinical and laboratory evidence suggests that cytokines have an etiologic role. Systemic corticosteroids are the therapeutic gold standard for Sweet's syndrome. After initiation of treatment with systemic corticosteroids, there is improvement of dermatosis-related symptoms and skin lesions. Topical application of high potency corticosteroids or intralesional corticosteroids may be efficacious for treating localized lesions. Other first-line oral systemic agents are potassium iodide and colchicine. Second-line oral systemic agents include indomethacin, clofazimine, cyclosporine and dapsone. The symptoms and lesions of Sweet's syndrome [22] may resolve spontaneously without any therapeutic intervention; however, recurrence may follow either spontaneous remission or therapy-induced clinical resolution.

Conclusion

Neck masses can be dicey and dubious. Each type of neck swelling has a unique location in the neck pertaining to its embryological origin. There must be one line that should be bored in the minds is that "not all neck masses are metastatic" and "not all neck swellings are carcinogenic". Neck swellings can sometimes have a multidisciplinary approach through Otorhinolaryngologists, Surgeons, Dermatologists, as clinical presentation of them are too vast and vague. There are a variety of fancy names associated with these neck masses making their diagnosis and treatment both massive and elusive. There can be a rarest of the neck swelling to commonest of the neck mass both appearing intricate and elaborate making the management of these masses in the neck picky and finicky.

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