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Book Review

Dermato-Fibro-Sarcoma and a Call for Global Attention to the High Cancer Deaths in Nigeria and Sub-Saharan Africa

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This article is one of the products of my response to the rare opportunity of going through Dermato-fibro-sarcoma protuberance (DFSP), a skin cancer I experienced on my head for about 30 years. From 1978, when I was just ten years old, till 2001, when I was about 33, various sizes of DFSP tumor grew in my scalp, which were removed through several dangerous surgical operations. Perhaps, I would not have needed to go through so many surgeries and traumas, had my case been diagnosed and discovered early as cancer. But until the fifth surgery, we had no idea what the terrible growths were, and none of those initial doctors took the tumors to the laboratory!

However, the prolonged, traumatic experience provided me a vista into some real challenges around cancer care in Nigeria and in many African countries. Few of these challenges are poor funding of research works on cancer, lack of effective cancer education, illiteracy (especially among the rural populations), and poverty. The result is the high cancer deaths in the continent. In Nigeria, for instance, the World Health Organization estimates that 4 out of five people with cancer eventually die of the disease!

The objective of this article, therefore, is not only to expound on DFSP but to draw global attention to the havoc it is wreaking in conjunction with other cancers in Africa, therefore calling for a better organized battle against the disease through strategic and sustained efforts.

Dermato-fibro-sarcoma protuberance (DFSP)

Generally, cancers are described by their association with specific parts of the anatomy, organs or body function (e. g. lung, bowel, leukemia, skin, breast, colon, pancreas, etc). And cancer has the ability to occur virtually on all organs in the body.

DFSP is one of the cancers people ignorantly live with and which are silently sending people to their early graves in sub-Saharan Africa. It is a type of cancer in the sarcoma family which is considered rare. Although there are 50 types of sarcoma, they can be grouped into two main categories: soft tissue sarcoma and bone sarcoma or osteosarcoma.

Description

Dermato-fibro-sarcoma protuberance (DFSP) is a low to intermediate grade soft-tissue sarcoma originating from the dermal layer of the skin. Although historically it has been attributed to fibroblastic origin, DFSP is now thought to originate from mesenchymal stem cells, according to the currently dominant cancer stem cell hypothesis. It was first described as "progressive and recurring dermatofibroma in 1924 by Darier and Ferrand", underscoring its predilection for local recurrence. DFSP is a locally aggressive tumor and despite sharing some histologic features with fibrohistiocytic tumors, it tends to grow in a more infiltrative manner. Three-dimensional reconstructions of DFSP have shown that the tumor can assume irregular shapes and extend in a villous or finger-like manner. These irregular, tentacle-like extensions are believed to be responsible for the common clinical dilemma of local recurrence following inadequate resection.

Epidemiology

This is the study about spread of diseases and their control. Although, information about this cancer is generally lacking among the people in Nigeria and many parts of Africa, it is generally believed that DFSP is not a common kind of cancer, but how rare is it?

According to study carried out by American Joint Committee on Cancer, DFSP comprises roughly .01% of all malignant tumors and approximately 2 to 6 percent of all soft-tissue sarcomas. The estimated incidence is 0.8 to 5 cases per 1 million persons per year, which translates to roughly 1,000 new cases per year in America. The incidence among blacks, which figure is 6.5 per million is almost double that among whites which is 3.9 per million. In most cases, Dermato-fibro-sarcoma affects patients between 20 and 50

years of age, even though it has been described in both children and in the elderly.

Clinical features

Dermato-fibro-sarcoma usually has a long slow sluggish course, with early tumors appearing as painless areas of cutaneous thickening. They may have pink, dark red or even bluish discoloration, particularly at the periphery. Over time, they develop into a larger nodular mass, and ultimately can develop into a large fungating lesion. When they grow into the epidermal layer of the skin, they may eventually ulcerate. Unlike tumors of the subcutaneous tissue, Dermato-fibro-sarcoma is adherent or intimate with its overlying skin.

DFSP's growth ranges from months to years. Often, it is erroneously taken for another type of sarcoma cancer called lipomas. At other times, it is mistaken for deep-seated epidermal cysts, scars, hypertrophic scars, keloids, dermatofibromas, nodular fasciitis, and insect bites and a delayed diagnosis is common, especially in places like Nigeria where there is generally poor awareness among the people.

The locations in human body where Dermato-fibro-sarcoma commonly occur are: the trunk, which is the most common location with (47%), followed by lower extremity (20%), upper extremity (18%), and finally head and neck (14%).

As it happened, my own DFSP case occurred in the scalp (the skin covering the skull), the rarest place that the disease occur in the body!

Metastases

This is the spread of cancer from its original locality to other organs or parts of the body. DFSP is a malignant tumor which has tendency for reoccurrence, but only metastasizes 1-4% of the time. Metastasis in DFSP is a late clinical outcome and typically occurs later in the disease's course or after several local recurrences.

Diagnosis of DFSP

The diagnosis of DFSP is basically done by oncologists. As early as the tumor is detected, effort must be made to contact medical experts who will carry out relevant diagnosis and advice on the appropriate steps to take towards treatment.

Some of the ways by which DFSP can be diagnosed include magnetic resonance imaging (MRI). This is used to evaluate the gross local extent of the tumor and may be important in preoperative planning for larger tumors. As with many other soft tissue tumors, T1-weighted images demonstrate low signal characteristics while T2-weighted images exhibit higher signal. While MRI can adequately delineate the bulk of the tumor, it does not define microscopic tumor extension. Furthermore, it does not clearly define recurrent lesions or lateral infiltration which is typical of this entity.

In patients with prolonged or recurrent DFSP or when sarcomatous changes are evident (DFSP-FS) a CT of the chest is obtained to evaluate for pulmonary metastases. A CT scan of the local area can be done, if bony involvement is suspected.

Diagnosis is made using either a core needle or an open incisional biopsy. While the role of fine needle aspiration is established in cases of recurrent disease, it is recommended that the initial biopsies be larger samples that demonstrate the histologic architecture of the tumor. Biopsy is the removal and examination of a sample of tissue from a living body for diagnostic purposes.

- **A core needle biopsy**: This is also known as core biopsy. It involves removal of a very small amount of tumor and is performed by inserting a hollow needle through the skin and into the organ or abnormality to be investigated. The needle is then advanced within the cell layers to remove a sample or core. This procedure takes a few minutes to perform and may be undertaken in an outpatient setting.

- **An incisional biopsy:** This is the removal of only a portion of the tumor for the pathologist to examine. A pathologist is an expert who examines samples of body tissues for diagnostic or forensic purposes. An incisional biopsy is generally reserved for tumors that are larger and offers the pathologist a larger specimen with which to work. This type of biopsy has a slightly higher diagnostic success rate and is usually carried out in the operating room.

- **An excisional biopsy**: This involves removal of the entire tumor and is typically reserved for very small lesions in which an incisional biopsy or a core needle biopsy is no longer applies. It is usually performed in cases where removing the entire lesion along with a narrow margin of normal tissue is easily accomplished and tolerated by the patient. This is also often performed in the operating room – as was my case during the fifth surgery in 2001.

Treatment

The medical options in the treatment of Dermato-fibro-sarcoma are solely the works of Oncologists and Dermatologists. Oncologists are doctors or scientists concerned with tumors, including the study of their development, diagnosis, treatment and prevention while Dermatologists are persons skilled in taking care of skin diseases.

The mainstay of treatment of DFSP has been surgery. Because of the high rates of recurrence, historical recommendations have sought 5 cm margins. Most cases of DFSP can be adequately treated by a dermatologist in an outpatient setting. However, in cases of very large or advanced DFSP, or where major reconstructive surgery will be needed, a multidisciplinary approach is used. This involves an oncologist, dermatologist, and a pathologist. In cases involving the deep tissues or bones, the participation of an orthopedic surgeon specializing in tumor surgery may be necessary. In cases where extensive surgical reconstruction will be necessary, a plastic surgeon may be called upon.

However, one highly effective procedure for the excision of DFSP and common types of skin cancers is Mohs surgery. Mohs Surgery, created by Dr. Fredrick E. Mohs, is microscopically controlled surgery and comes with complete excision with microscopic margins. This procedure has yielded excellent outcomes and offers the benefit of decreased surgical morbidity. In a comparative study of wide resection versus Mohs surgery, wide resection was associated with a recurrence rate of 13% whereas Mohs surgery had no recurrences at 5 years. Evidence is accumulating that Mohs surgery is the optimal surgical choice for most DFSP lesions with the exceptions of the aforementioned large/advanced DFSP that may require more extensive surgical and reconstructive procedures.

The Mohs surgery involves four steps which are: 1. Surgical removal of tissue 2. Mapping the piece of tissue, freezing and cutting the tissue, and staining with H&E or other stains 3. The interpretation of microscope slides and 4. The reconstruction of the surgical defect

The procedure is usually performed in a physician's office under local anesthetic. A small scalpel is utilized to cut around the visible tumor. A very small surgical margin is utilized, usually with 1 to 1.5 mm of "free margin" or uninvolved skin. Because the Mohs procedure is microscopically controlled, it provides precise removal of tumor, while healthy tissue is spared.

Other treatment procedures of DFSP include chemotherapy and the use of radiotherapy

Chemotherapy is the use of chemical treatment to kill or halt the replication and/or spread of cancerous cells in a patient. Radiotherapy, on the other hand, is the use of ionizing radiation, almost exclusively used for the treatment of malignant disease. DFSP is considered a radio-sensitive tumor.

Prognosis

This is the prediction or forecast of the course of a disease. When a case of DFSP is presented early enough, the general prognosis is excellent. The overall rate of distant metastasis is only 5% and regional metastasis is 1%. Historically, recurrence rates have been high, ranging from 11%- 53%, but with the advent of Mohs surgery, the rates have dropped. Even with recurrent DFSP, Mohs surgery has a 98% cure rate.

Conclusion

When DFSP or any cancer type is detected early and appropriate treatment options applied, many people can experience life after cancer. However, as a cancer survivor and an active cancer advocate in Nigeria, I am very familiar with state of cancer care in the country.

The complete absence of oncological services at the primary healthcare level, lack of good spread of cancer diagnostic centers, lack of sufficient facilities at the secondary and tertiary health facilities, inadequate manpower in the oncology sector, lack of sustained cancer awareness, coupled with widespread poverty are some of the dynamics for high morbidity – why 4 out five people with cancer eventually die in the country! These factors, no doubt, have brought Nigeria, and indeed Africa, under the siege and bondage of cancer. The time has come that the world must pay attention to the ravaging cancer war going on in Nigeria and the Sub-Saharan Africa. The current dangerous trend present opportunities for both African governments, international agencies and other critical stakeholders in the cancer care world to rise now to fight and conquer the seemingly invincible, silent killer disease.

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