Original Article

A Novel Approach in the Diagnosis of Pseudoxanthomaelasticum by Flame Test: Advocating the Payal-Shakil Phenomenon

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Abstract

Background: Pseudoxanthomaelasticum (PXE) is an uncommon dermatological disorder characterized by elastic fiber degeneration and calcifications. These calcified structures bear much importance in the diagnosis of the disease. **Aim:** To determine the nature of calcification present in biopsied skin lesions of PXE by using flame test to contribute in the diagnosis. **Materials and Methods:** Flame test of the hard calcified mass was carried out maintaining the standard laboratory protocols. A yellow- red color was seen at the Clear zone of the Bunsen burner flame. **Results:** Interpretation of the identified yellow- red color flame was done based on color indicators from the standard chart for flame test identification. The compound was confirmed to be calcium. **Conclusion:** Flame test is used for the first time in diagnosis of calcifications in medical disorders and we call this novel approach as the Payal-Shakil phenomenon highlighting the need to make use of every detail available of the specimen which is valuable in making correct diagnosis.

Keywords: Pseudoxanthoma Elasticum, Flame Test, Payal-Shakil phenomenon

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Introduction

Pseudoxanthomaelasticum (PXE) is a rare hereditary connective tissue disorder, characterized by generalized elastic fiber degeneration, first described in 1881 by Rigal. The clinical features of this disorder consists mainly of cutaneous, ocular, and vascular manifestations; comprising the "Gronblad-Strandberg syndrome". Skin histopathology involves swollen, degenerated irregularly clumped and fragmented elastic fibers in the middle and deep dermal layers, with secondary calcium deposition in the fibers.[1-4]

The cutaneous lesions are small yellowish papules or larger coalescent

plaques that gives a cobblestone appearance to the skin.[1,2] The affected skin results in hanging and redundant folds when it gets severe. Skin lesions are seen mainly at areas of flexion, such as the neck, axillae, antecubital and popliteal fossae, inguinal areas, and periumbilical region.[5] The ocular manifestations are seen as "angioid streaks"that are the characteristic funduscopic findings, caused by calcification of the elastic fibres of the Brusch membrane of the retina and may ultimately lead to cracking and fissuring with retinal hemorrhages, scarring and central vision.[6-8] The vascular manifestations in PXE are caused by degeneration of the elastic lamina of the arterial wall, often with calcium deposition.[9] Neurological complications are rare which have been reported by many authors.[10]

The biopsied specimen of PXE is often accompanied with various calcifications. In this present case of ours, we wanted to know the nature of hard tissue present in the specimen by performing a Flame Test. The specimen was also verified by the use of Stereo-microscopy to be a calcified structure. (Figure-1)

Method:

Flame Test

It is a qualitative test used to visually know the identity of a metal or metalloid based on the characteristic color the salt turns the flame of a Bunsen burner. The heat of the flame excites the electrons of the metal ions causing them to emit visible light. Every element has a signature emission spectrum that is used to differentiate one element from another.

The biopsied specimen sent for histopathological examination contained presence of calcified mass (Figure 2). A nickel-chromium wire loop was cleaned by dipping it in hydrochloric acid followed by rinsing in distilled water. The cleanliness of the loop was tested by inserting it into a gas flame. After ensuring the cleanliness of the loop the flame test was carried out. The loop carrying the sample was placed in to the clear or blue portion of the flame and the emission of the characteristic yellow-red color was noted confirming the presence of calcium compounds in the specimen obtained. (Figure 3, Figure 4 and Table-1)

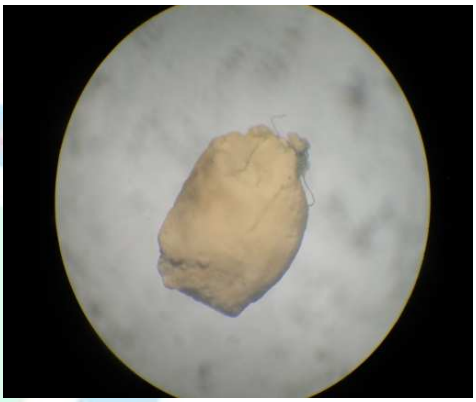

Figure 1: Stereo-microscopy Of The Calcified Mass

Figure 2: Gross appearance of Tissue specimen

Figure 3: Loop carrying the specimen into the blue part / clear zone of the bunsen burner.

Figure 4: Yellow- red color flame seen on burning of the specimen indicating presence of calcium ions

Figure-5: Tissue specimen demonstrating black calcium deposits on special staining with Von- kossa (10X Magnification)

Results

The sample is identified by comparing the visible spectrum of flame color against known values from a table or chart (Table 1)

Table 1: Interpretation Of flame test

Color	Metal Ion
Red	Carmine to Magenta:- Lithium compounds Scarlet or Crimson:- Strontium compounds Red-:Rubidium
Yellow-Red	Yellow-Red:- Calcium compounds
Yellow	Gold:- Iron Intense Yellow:-Sodium compounds
Blue	Light Blue:-Arsenic Azure:- Copper compounds (moistened with HCl)
Green	Emerald:-Copper compounds Bright Green:- Boron Blue-Green :- Phosphates (moistened with H_2SO4) Yellow-Green:- Barium
White	Bright White:- Magnesium White-Green :-Zinc
Purple	Violet:- Potassium compounds

Discussion

Calcium is very vital to many biologic processes. It plays a very important role in keratinocyte proliferation, differentiation and cell-cell adhesion. Serum calcium is tightly regulated by parathyroid hormone and 1,25(OH)2D3. The presence of calcium calcifications in pseudoxanthomaelasticum becomes the

most characteristic feature for its diagnosis.[11]

An observation of this kind is made for the first time wherein application of flame test is advocated in determining the nature of calcified compounds present in PXE.

We call this as a *Payal-Shakil* phenomenon and this novel approach can be used in diagnosis of various dermatological conditions and other calcifications present in different diseases. It is not very uncommon to encounter several biopsied lesions with hard calcifications in the pathology laboratory. Techniques such as ground sectioning, X-Ray crystallography etc. have been used to demonstrate the 3-dimensional structure of the calcifications. Flame test is very easy to perform and enables one to concise the differential diagnosis and can be a first step in further final diagnosis. In the present case after the initial confirmation of calcium compound in the tissue specimen by flame test (*Payal-Shakil* phenomenon) we carried out a special staining with Von-Kossa stain later (Figure-5), to confirm the presence of calcium.

Conclusion

Every attempt should be made to make the best use of all the bits of specimen available using biochemical and molecular techniques for early and prompt diagnosis of various disorders. The current phenomenon can be performed in various diseases and the inference can be drawn based on presence/absence of the chemical compounds for a rapid and easy diagnosis.

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